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Clinical and Radiological Effect of Epidural Blood Patch in a Patient with Marfan Syndrome, Chiari I Malformation, Intracranial Hypertension, and Dural Ectasia: Case Report and Systematic Review

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Abstract

Background and Purpose— Patients with Marfan syndrome can develop headaches and upright posture intolerance due to dural ectasia and/or cerebrospinal fluid (CSF) leakage. The role of epidural blood patch in such patients remains unclear.

Methods— We present a case of a 26 year-old woman with Marfan syndrome, hydrocephalus, sacral dural ectasia, and tonsillar herniation, presenting with headache and upright posture intolerance. Cerebrospinal fluid pressure monitoring demonstrated elevated pressure in supine and upright positions with no evidence of a CSF leak on computed tomography (CT) myelogram. Two epidural injections of autologous blood and fibrin were performed 5 months apart using caudal and lumbar approaches, respectively. A systematic review of the literature was performed to determine outcomes of epidural blood patch in patients with Marfan syndrome.

Results— After the first epidural blood patch, the patient had marked clinical improvement that persisted for 1 month and partial improvement that persisted for 2 months. After the second epidural blood patch, the patient had marked clinical improvement that persisted for 2 months and partial improvement that persisted from last follow-up to 18 months. The tonsillar descent was measured at 10 mm at baseline evaluation and demonstrated reduction in magnitude of descent after epidural blood patch; 2.3 mm and 3 mm reduction after first and second epidural blood patches, respectively. A total of 9 patients with Marfan syndrome who were treated a total of twelve epidural blood patches (nine performed in the lumbar region, two in the thoracic region, and one in the sacral region) were identified. The follow-up period ranged from 3-months to 18-months post-treatment. Of the seven patients with long-term follow up, complete resolution of symptoms was seen in four and partial resolution of symptoms seen in three patients.

Conclusions— Our report suggests that patients with Chiari malformation and dural ectasia can experience clinical and radiological improvement even without evidence of intracranial hypotension or CSF leak.

Keywords— Dural ectasia, tonsillar herniation, Chiari malformation, Marfan syndrome, epidural blood patch, fibrin injection, hydrocephalus.

INTRODUCTION

Epidural blood patches have been attempted in acquired Chiari malformation because of clinical benefit and reversal of tonsillar descent observed in patients with intracranial hypotension with CSF leakage. Isolated reports of short-term results of epidural blood patch in patient with Marfan syndrome and Chiari 1 malformation have been published.[1-4] Almost all the reports identify intracranial hypotension due to CSF leakage in these patients due to inherent weakness of dura. Inherent weakness of dura also leads to dehiscence in the lumbosacral region leading to dural ectasia in Marfan syndrome.[5-8] The mechanisms of action of epidural blood patch is thought to be due to sealing of leakage site and reducing epidural space compliance and increase in local cerebrospinal fluid pressure in lumbar region and reducing the caudal movement of cerebellar tonsils. The case reports are based

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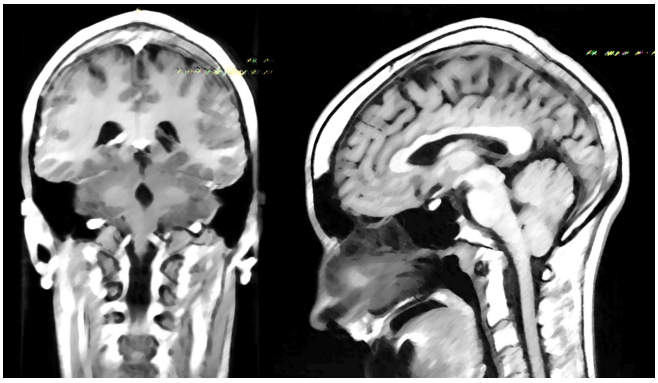


FIGURE 1: MRI brain demonstrating dilation of lateral and fourth ventricle (coronal view) and cerebellar tonsillar herniation (sagittal view).

on short-term follow-up with unknown long-term results. We present a case of Marfan syndrome with hydrocephalus, intracranial hypertension, Chiari malformation, and dural ectasia who presented with upright posture intolerance and postural headaches. We report the long-term clinical and radiological outcomes following epidural injection of blood and fibrin sealant.

CASE DESCRIPTION

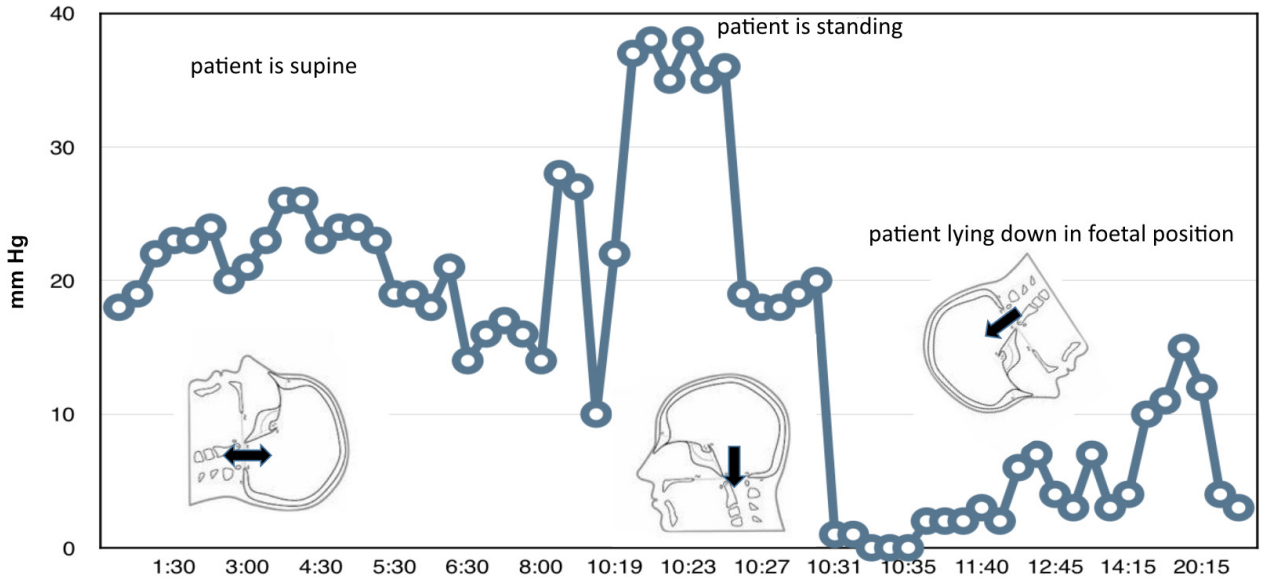
Clinical history

A 26-year-old woman with past medical history of Marfan syndrome and epidural ectasia presented with severe headaches for last 4 years. Initially, the headache was sudden in onset, ranging in severity between 4 and 5 (numeric rating scale), located at occipital region with radiation to the neck. The headaches were associated with neck stiffness, and were aggravated by standing upright and relieved in fetal (extreme flexion) position. The patient had tried several medications such as riboflavin, magnesium, prednisone, amitriptyline, sumatriptan, topiramate, toradol, cyclobenzaprine and carisoprodol, all with limited benefit. The severity of headaches increased and dihydoergotamine was initiated, but also provided no relief. The patient had previously undergone an epidural blood patch with temporary partial relief. One year prior, the patient underwent surgery for repair of an aortic aneurysm. Patient reported occurrence pre-chiasmatic episodic visual changes later in the year. The patient had also undergone eye surgeries six years prior. Magnetic resonance imaging (MRI) of the brain demonstrated hydrocephalus and cerebellar tonsillar descent (see Figure 1). MRI of the spine demonstrated dural ectasia at sacral level and peri-neurial cysts at lumbar 2 and 3 vertebral level. There was cerebellar tonsillar descent of 10 mm measured on MRI.

Cerebrospinal fluid pressure measurements

There was ambiguity regarding intracranial and CSF pressures as either intracranial hypotension or hypertension may be associated with Chiari malformation and hydrocephalus and perineural cysts are more common in those with intracranial hypertension.[9-10] CSF pressure monitoring was considered valuable prior to performing another epidural blood patch. CSF pressure monitoring was performed using a closed tip lumbar catheter (Integra Neurosciences, Plainsboro, NJ, 80 cm, 5 Fr) inserted between the spinous processes and laminae of lumbar 4 and 5 vertebrae. The lumbar catheter was connected to a pressure monitoring system with transducer positioned at the level of the Foramen of Monro.[11] The monitoring system was re-calibrated whenever there was a change in the elevation of the head of the bed relative to the pressure transducer. We also acquired CSF pressure recordings in upright position by placing the transducer at level of distal end of inserted lumbar catheter (at thoracic 10 vertebral level). Figure 2 demonstrates the representative CSF pressure throughout the two days of monitoring. The CSF pressure ranged from 18 to 28 mm Hg with mean of 22.2 mm Hg during lying position (measured 16 times). CSF pressures were elevated in most of the recordings being highest in standing position (range 23 - 40 mm Hg with a mean of 38.7 mm Hg, measured 10 times) and lowest in extremely flexed posture (range 0-2 mm Hg with a mean of 0.5 mm Hg) measured 4 times. A total of 55 cc of CSF was drained in 20 hours as a trial. Patient reported improvement in

FIGURE 2: Cerebrospinal fluid pressure measurements at different body positions.



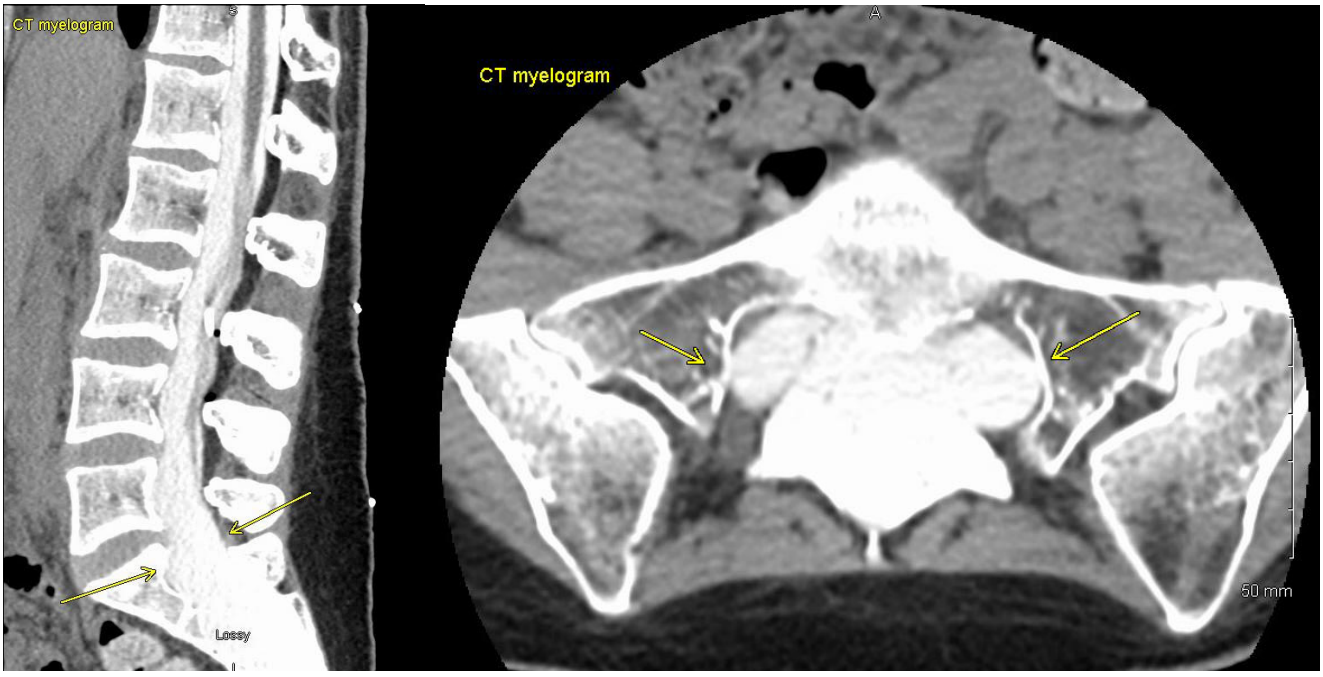


FIGURE 3: CT myelogram demonstrating dural ectasia in sacral region.

back pain and its radiation after CSF drainage. A myelogram was performed prior to removal of lumbar catheter which demonstrated ectasia of cerebrospinal space in the lumbar region (see Figure 3). There was dilatation of subarachnoid sac in the sacral region with perineural sheath dilation. There was no CSF leak identified.

Epidural blood and fibrin injection

Despite high CSF pressures, an epidural blood injection was considered to reduce cerebellar tonsillar descent. Patient underwent epidural blood patch and fibrin sealant injection after 1 month following CSF pressure monitoring, using a caudal sacral approach. The procedure was performed under awake conditions. An 18-gauge 3-inch spinal needle was advanced between the sacral hiatus above the coccygeal bone. A 3-cc syringe was attached to spinal needle with gentle pressure to identify the epidural space by loss of resistance method. Approximately 3 cc of radio-opaque contrast was injected to confirm the cephalad posterior epidural spread on the lateral view. A total of 15 cc of autologous venous blood was injected into the epidural space followed by a total of 8 cc fibrin sealant Tisseel (Baxter, Deerfield, Illinois) (see Figure 4). The patient had significant clinical improvement for 2 weeks but then presented with recurrence of headache which was manageable with increased dose of fentanyl and caffeine-based products. Patient had been able to stand up and ambulate. The follow-up MRI demonstrated reduction in tonsillar descent to 7.7 mm. She was subsequently sent for neurosurgical consultation for surgical decompression, but she was not found to be eligible candidate for the any neurosurgical procedure.

Patient underwent another epidural blood patch and fibrin injection 5 months later. An 18-gauge 3-inch spinal needle was advanced into left lumbar 3 and 4 vertebrae interspace using paramedian approach. A 3-cc syringe was attached to spinal needle with gentle pressure to identify the epidural space by loss of resistance method. Approximately 3 cc of radio-opaque contrast was injected to confirm the cephalad posterior epidural spread on the lateral view. A total of 10 cc of autologous venous blood was injected into the epidural space, followed by a total of 22 cc of fibrin sealant Tisseel (Baxter). Another 10 cc of autologous venous blood was injected after the injection of fibrin sealant into the epidural space. The patient reported immediate improvement in level of awareness, ability to stand and upright tolerance from 3-5 minutes to 90 minutes' post procedure and showed

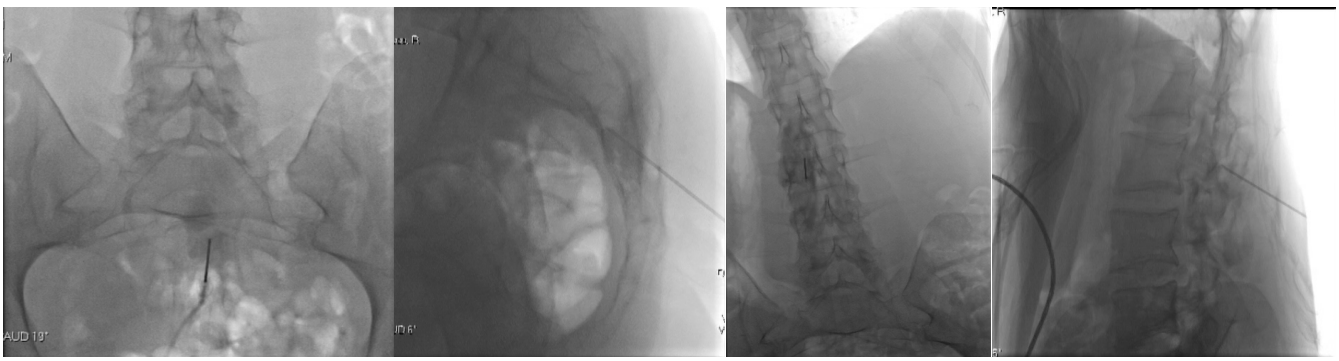


FIGURE 4: Radiographic images demonstrating epidural injections of autologous blood and fibrin were performed 5 months apart using caudal and lumbar approaches, respectively.

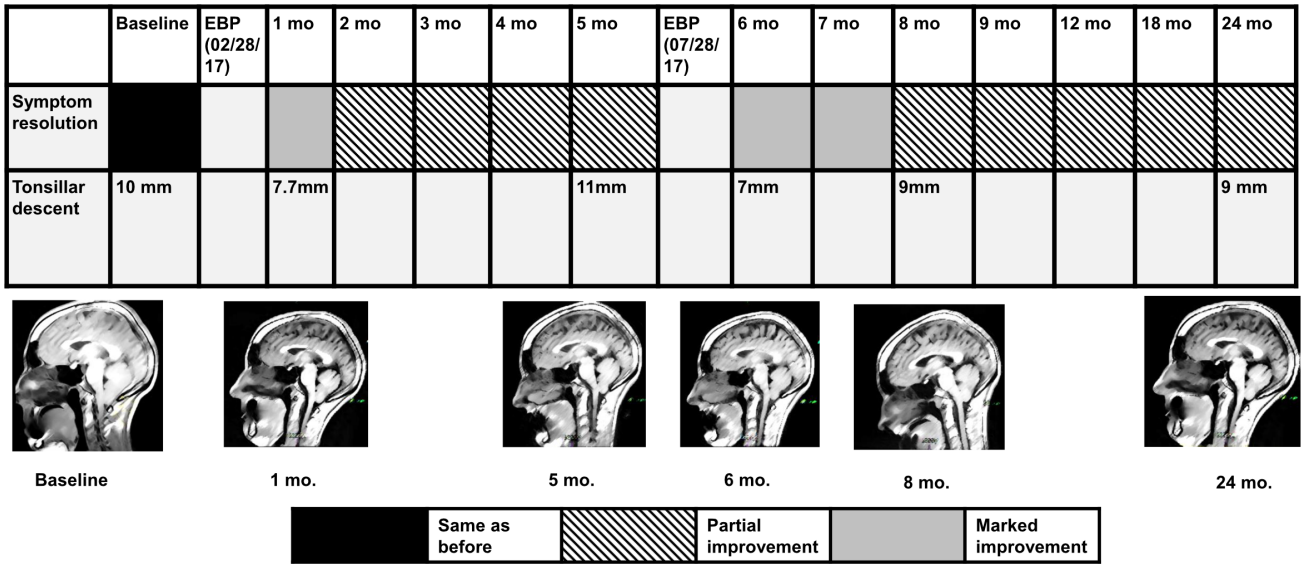


FIGURE 5: Serial clinical and radiographic assessments over a 24 month period.

improvement in quality of life. At 4-month follow up, she demonstrated continued clinical improvement in headaches and was able to tolerate upright posture for 2-3 hours.

Assessment of serial clinical and radiological changes

We analyzed all the MRI images available to us (six MRIs over 3 years). We measured tonsillar descent on T1 -weighted spin-echo images according to the method described by Barkovich et al.[12] The measurements were made from the lower level of the foramen magnum to the bottom of the lowest lying tonsil. The bottom of the foramen magnum was defined as a line extending from the lowest cortical bone seen at the posterior lip of the foramen magnum to the lowest cortical bone of the clivus. The tonsillar descent was measured at 10 mm at baseline evaluation and demonstrated reduction in magnitude of descent after epidural blood patch; 2.3 mm and 3 mm reduction after first and second epidural blood patches, respectively (see Figure 5). The tonsillar decent was 2 mm less at 18 months post-procedure after second epidural blood and fibrin sealant injection compared with pre-procedure measurements.

Patient's severity of symptomology was determined using evaluations made by neurologists at clinic visits. The severity of symptoms was classified in four grades using the findings at baseline visit prior to lumbar catheter placement and epidural blood patch treatments: 1/. Worse than before; 2/. Same as before; 3/. Partial improvement; and 4/. Marked improvement. The determination was based on documentation regarding severity of headache and upright posture intolerance.[13] Patient demonstrated marked improvement after each epidural blood patch. After the first epidural blood patch, patient had marked clinical improvement that persisted 1 month and partial improvement that persisted for 2 months. After the second epidural blood patch, patient had marked clinical improvement that persisted 2 months and partial improvement that persisted at last follow up to 18 months.

SYSTEMATIC REVIEW

Literature Search

For the purpose of identifying cases in which an epidural blood patch was performed in a patient with Marfan syndrome, PubMed was searched up to December 1st, 2019 using the key terms "Marfan syndrome", Marfan's syndrome", "epidural blood patch", "blood patch", "dural leak", "cerebrospinal fluid leak", "spontaneous spinal cerebrospinal fluid leak", "intracranial hypotension", "spontaneous intracranial hypotension", "postural hypotension", "headache", "postural headache", "post dural puncture headache", "tonsillar herniation", "Chiari malformation" (Table 1). Cross-references and related articles identified additional studies that may have been overlooked in the initial search.

Data Collection

For each included study, the following data was extracted: (i) clinical parameters for study: clinical attributes on presentation, spinal features attributed to Marfan syndrome, presence or absence of Chiari malformation, demonstration of cerebrospinal fluid leak; (ii) procedural details: level of epidural blood patch, number of epidural blood patches performed, intra-procedural complications; (iii) study outcomes: clinical improvement post-procedure and on long-term follow-up, long-term complications. The main primary outcome was the level of improvement in clinical symptoms immediately post-procedure and on long-term follow-up.

Results of Systematic Review

A total of 9 patients with Marfan syndrome presenting with headache consistent that was treated with an epidural blood patch were identified (Table 1). The mean age was 23.3 years (±10.8) and 7 were women (77.8%). Chiari 1 malformation was seen in two of the nine patients and CSF leak was documented in six patients. A single epidural blood patch was performed in six patients and multiple epidural blood patches were performed in three patients. There was a total of twelve epidural blood patches performed, across the nine

TABLE 1: Search terms utilized in PubMed.

	PubMed Search
1	((intracranial hypotension) or (spontaneous intracranial hypotension) or (SIH) or (postural hypotension))
2	((headache) or (postural headache) or (post dural puncture headache) or (PDPH))
3	((dural leak) or (cerebrospinal fluid leak) or (CSF leak) or (spontaneous spinal cerebrospinal fluid leak) or (spontaneous CSF leak))
4	((Marfan syndrome) or (Marfan's syndrome) or (Chiari malformation) or (tonsillar herniation))
5	((blood patch) or (epidural blood patch) or (lumbar blood patch) or (lumbar epidural blood patch) or (cervical blood patch) or (cervical epidural blood patch))
6	(search 1) and (search 2) and (search 3) and (search 4) and (search 5)

Abbreviations used:

SIH	spontaneous intracranial hypotension
PDPH	post-dural puncture headache
CSF	cerebrospinal fluid

studies, with nine performed in the lumbar region, two in the thoracic region, and one in the sacral region. The follow-up period ranged from 3-months to 18-months post-treatment. In two cases, no long-term follow-up was available. All patients experienced improvement in the severity of symptoms post-epidural blood patch treatment, especially in the immediate post-procedural period. Of the patients that were followed up long-term, complete resolution of symptoms was seen in four and partial resolution of symptoms seen in three patients. In one patient with coexisting Chiari 1 malformation, a reduction of 2 mm in tonsillar descent was seen on imaging at latest follow-up (18 months).

DISCUSSION

We report a patient with Marfan syndrome with Chiari 1 malformation, hydrocephalus, dural ectasia, and elevated CSF pressure. There were considerable positional changes in CSF pressures. There was improvement in clinical symptoms with low volume removal of CSF further supporting the presence of high CSF pressure contributing to the clinical manifestations. The mechanism for increase in CSF pressure and worsening of clinical symptoms are best explained by downward shift and increase in descent of cerebellar tonsils during upright position leading to impaction and compression of CSF pathways.[10] Epidural injection of a mixture of blood and fibrin sealant led to clinical improvement in symptoms and radiological improvement in tonsillar descent. Epidural injection using caudal approach and standard lumbar approach provided short-term benefit, but the patient reached a point of stability after the second epidural blood and fibrin injection performed through the lumbar approach. The benefit is probably related to reducing epidural space compliance and increase in local CSF pressure in lumbar region and reducing the caudal movement of cerebellar tonsils during upright posture.[14-15] Multiple injections in the epidural space probably resulted in fibrosis and reduced compliance within the dura leading to a sustained benefit.[16-17]

Previous investigators have reported the results of epidural blood patch in patients with Marfan syndrome with or without cerebellar tonsillar descent. Unlike our patient, many of the patients had documented low CSF pressure and CSF leak. Puget et al.[1] reported a 12-year-old woman with Marfan syndrome, sacral dural ectasia, and cerebellar tonsillar herniation on MRI. The patient had presented with 6-month history of biparietal and occipital headaches, most severe in an upright position, and relieved in supine position. Patient underwent a foramen magnum decompression with opening of cervical 1 lamina and durotomy. Intra-operatively, there was a paucity of CSF with relative collapse of subarachnoid space noticed after dura exposure suggesting intracranial hypotension. Due to lack of clinical improvement, MR myelography was performed which confirmed the presence of sacral meningeal diverticula and an ongoing CSF leak on the left side at the level of the S1 nerve root. The patient was treated with an autologous epidural blood patch at the L5–S1 level resulting in prominent initial transient improvement. One more blood patch was performed on day 8 resulting in persistent resolution of symptoms for next 15 months. Milledge et al.[3] reported upon a 14-year-old girl with a 6-week history of holocranial headache which were most severe in an upright position and relieved in supine position. Cerebellar tonsillar herniation was detected on MRI. Surgical decompression of the foramen magnum was performed with no improvement in clinical symptoms. A spinal MRI scan demonstrated lumbosacral dural ectasia and perineural cysts around the sacral nerve roots. There was no spontaneous drainage of CSF when lumbar puncture was performed, confirming that the CSF pressure was low although no CSF leak could be identified. Patient underwent epidural blood patch at L2−3 with partial relief from her symptoms. She remained pain free over the subsequent 12 months. This patient presented again to a regional hospital with similar headaches 20 months after her initial presentation. The patient had another epidural blood injection resulting in modest reduction in severity of headaches and required long-term oral analgesia. Roster et al[18] reported upon a 14-year-old female with Marfan syndrome who presented with headache for a month which were most severe in an upright position and relieved in supine position. Brain MRI with gadolinium demonstrated diffuse pachymeningeal gadolinium enhancement, bilateral subdural hematoma, decreased size of ventricles, cerebellar tonsillar herniation, and enlarged pituitary gland. MRI myelography showed dural ectasia with meningocele, large radicular cysts and large lumbosacral arachnoid diverticula without any CSF leak. Autologous epidural blood injection was performed at lumbar 4 and 5 vertebral level. One month after the blood patch, patient had complete resolution of headache and returned to school full time. Three months after the epidural blood patch, there was improvement in cerebellar tonsillar herniation on repeat MRI. One year later, the patient remained asymptomatic, and brain MRI was normal. Pabaney et al.[4] reported upon a 38-year-old man with Marfan syndrome who presented with orthostatic headaches and seizures. The patient developed bilateral neck pain, which progressed to a holocranial headache which would get markedly worse on sitting, standing or bending forward and relived by lying down over a period of 2 weeks. MRI revealed pachymeningitis, atlanto-axial subluxation, cerebellar tonsillar herniation and flattening of pontine surface. MR myelogram demonstrated dural ectasia and expansion of thecal sac, more marked in

TABLE 2: Summary of existing literature on patients with Marfan syndrome receiving epidural blood patch.

Author(s)	Year	Clinical attributes	Marfan syndrome attributes	Chiari malformation	Demonstrated CSF leak	Epidural blood patch level	Number of epidural blood patches performed	Short-term outcomes	Long-term outcomes	Reduction of tonsillar descent (if Chiari malformation present)
Davenport, et al	1995	Right-sided throbbing headache exacerbated by sitting up for past 2 years. Associated with nausea, vomiting	-Lumbar arachnoid diverticula around lumbrosacral nerve roots	N	Y	T1-T3	2	Immediate improvement in symptoms (pain analogue score of 1-2 out of 10)	6 weeks post-first epidural blood patch, headache returned with less intensity. Repeat thoracic epidural blood patch performed: pain improved (pain analogue score of 2-4 out of 10) 3 months post-repeat blood patch procedure	Not applicable
Fukutake, et al	1996	Occipito-nuchal headaches exacerbated upon standing and walking for past 15 years	-Lumbrosacral arachnoid diverticula around lumbrosacral nerve roots	N	Y	L4-L5	1	Immediate improvement in symptoms (pain analogue score of 10 at presentation to 5 post-treatment)	No long-term follow-up available	Not applicable
Milledge, et al	2005	Holocranial headache for 6 weeks. Exacerbated on standing/sitting.	-Dural ectasia (lumbrosacral)-Posterior scalloping of lumbar vertebrae-Tarlov cysts around sacral nerve roots	Y	N	L2-L3	1	Pain free 24 hours post-treatment	Pain free over 12 months post-treatment.	Not documented
Rosser, et al	2005	Bifrontal headache for 3 weeks. Exacerbated on standing upright, and associated with nausea, vomiting, and photophobia	-Dural ectasia (multiple areas)	N	Y	L5	1	Partial improvement: moderately painful bifrontal headaches 6 weeks post-treatment	Pain free 4 months post-treatment	Not applicable
Puget, et al	2007	Headache: biparietal and occipital. Exacerbated in upright position and refractory to analgesic treatment	-Dural ectasia (sacral)	N	Y	L5-S1	2 (2nd done 8 days after first blood patch)	Marked improvement over 5 days post-treatment	No evidence of recurrence or complication at 15 months post-treatment	Not applicable
Albayram, et al	2008	Headache exacerbated on standing.	-Meningeal cysts (multiple)	N	Y	L4-L5	1	Headache free 2 days post-treatment	No complaints or neurological findings at 3 months post-treatment	Not applicable
Pabaney, et al	2010	Holocranial headache exacerbated on sitting, standing, or bending forward for past 2 weeks, seizure episode	-Dural ectasia (multiple areas)	N	Y	L4-L5	1	Near complete resolution of symptoms 96 hours post-treatment	No evidence of recurrence or complications at 6 months post-treatment	Not applicable
Khalid, et al	2012	Positional headache for past 15 days with neck stiffness and a seizure episode	-Dural ectasia (sacral)	N	N	L4-L5	1	Moderate symptomatic improvement immediately post-treatment; Complete pain relief 48 hours post-treatment	No long-term follow-up available	Not applicable
Qureshi, et al (current case report)	2020	Occipital headache for the past 4 years. Exacerbated on standing position.	-Dural ectasia at sacral level -Perineural cysts at L2-L3	Y	N	Sacral (1st blood patch) L3-L4 (2nd blood patch)	2	After 2nd epidural blood patch: marked clinical improvement for 2 months	Partial improvement until last follow-up 18 months post-treatment	2.3 mm after 1st blood patch and 3 mm after 2nd blood patch (10 mm at baseline). At 18 month follow-up: reduction by 2 mm

the lumbosacral region with multiple dilated out-pouching in the sacral regions, S1-3. Accumulation of fluid in the posterior para-spinal muscles was also observed suggestive of CSF leaks. An epidural blood injection at lumbar 4 and 5 vertebral level with 25 ml of autologous blood resulted in marked improvement and almost complete resolution of his symptoms over the next 96 hours. He was followed up regularly as an outpatient for the next 6 months with no evidence of any complications or recurrence and returned to normal baseline activity and lifestyle.

Epidural blood injections have been reported in patients with intracranial hypotension and CSF leaks without Chiari malformation. Fukutake et al.[19] reported upon a 30-year-old woman with Marfan syndrome had chronic intractable headaches and spontaneous intracranial hypotension (CSF pressure 6 cm H20) without Chiari malformation. The pain was concentrated over the occipitonuchal region and was often aggravated by upright posture. CT myelography showed multiple, large, lumbosacral arachnoid diverticula and radioisotope cisternography indicated CSF leakage. Epidural blood injection resulted in immediate relief from the positional headaches. Davenport et al.[20] reported upon a 19-year woman with a six-week history of worsening headache concentrated over the right side of the head and throbbing in nature. There was worsening of headache with upright posture and spontaneous intracranial hypotension (CSF pressure could not be measured) without Chiari malformation on MRI. Radioisotope cisternography showed an abnormal accumulation of isotope on the left at the level of T1-3 with multiple smaller bilateral projections at most levels of the thoracic spine. Computed tomographic myelography disclosed contrast extruding from the left neural foramen at TI with a small amount of contrast passing medially to the pleura. In the lumbar region there were multiple large arachnoid diverticula around the lumbrosacral nerve roots with some erosion of the sacrum. After a lumbar epidural blood injection with no response, another epidural blood injection was performed at thoracic level resulted in considerable improvement of her symptoms. Patient was able to ambulate without restriction for the first time in several weeks. The symptoms recurred after 6 weeks requiring a second thoracic epidural blood injection. A subsequent radioisotope cisternogram, identified no residual CSF leak. Three months after the second thoracic epidural patch, patient had partial relief from headaches requiring regular analgesia but was able to return to work. Albayram et al.[2] reported upon a 32-year-old woman with a chronic headaches and polycystic kidney. The headaches were most severe in an upright position and relieved in supine position. There was no spontaneous drainage of CSF on lumbar puncture, confirming that the CSF pressure was below atmospheric pressure. Loss of dural sac integrity and dural leakages, contrast material extravasation into epidural area, and paravertebral region were detected on MR cisternography at lumbar levels and multiple meningeal cysts were identified at the dorsal area. Because patient's symptoms persisted despite conservative treatment for 6 weeks, epidural blood injection was performed. Two days later, the headache was fully recovered, and she was able to carry out her daily activities. Three months after the treatment, there were no complaints or neurological findings.

Our report suggests that patients with Chiari 1 malformation and dural ectasia can experience clinical and radiological improvement even without intracranial hypotension or CSF leak. Multiple epidural blood patches with fibrin sealant may be required. New agents such as NBCA[21-22] may increase the duration of benefit although further studies are warranted.

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