Metameric thoracic lesion: report of a rare case and a guide to management

Case report

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Metameric lesions of the spine are rare. The authors present a case of patient with a complex metameric vascular lesion of the thoracic spine and describe a management strategy for this entity.

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In the spine, a metamere is defined as the vertebral disc and half of the body above and below that disc. A metameric lesion is defined as a vascular lesion that is genetic and nonfamilial,1 is extramedullary and extradural, and can involve bone, soft tissue, and muscle.2 In an extreme form, Cobb syndrome, involvement extends to the skin.3,5,6,9 Rodesch and Lasjaunias1 theorized that the earlier such a lesion forms in the embryological timeline, the more extensive a lesion will tend to be, stating “This is due to the fact that endothelial cells of the spinal cord share the same mesodermal origin as skin or muscles.”2,9

Metameric lesions of the spine are very rare with only a few reported cases in the literature and little information on how to manage these complex lesions.5,9 However, some authors believe that these lesions are underreported, especially in the pediatric population.3 Ten to 21% of pediatric patients with spinal arteriovenous shunts may actually demonstrate a metameric distribution to their lesion.4,8 Additionally, patients with multiple lesions not fitting an exact metameric pattern but otherwise appear metameric may be more aptly classified as metameric with variable expression.3

We report a case of a patient with a large metameric-like lesion and describe our management strategy, outcome, and suggestions for treatment of similar lesions.

Case Report

History and Examination. This 17-year-old girl was noted at birth to have a subcutaneous lesion on her back. She presented to her pediatrician with a 2-week history of progressive gait difficulty. She denied any bowel or bladder difficulty. In the past, she had been told that the lesion on her back was a large lipoma and did not require any treatment, and no imaging was done.

On examination, she had muscle strengths of 4/5, 4+/5, and 4/5 in her right iliopsoas, right quadriceps, and right ankle dorsi- and plantar flexion, respectively. She had markedly decreased proprioception in her right great toe and a positive Babinski sign on the right. She was not able to stand on her own, but with assistance she had a spastic, scissoring gait. Otherwise, she was neurologically intact. On her back, there was a large, soft, immobile mass extending from the base of her cervical spine to the top of her lumbar spine with no overlying skin lesion or changes in skin pigment. She had no other neurocutaneous lesions. Funduscopic examination findings were normal.
Magnetic resonance imaging and CT scanning studies were performed and revealed a very large, complex mass mostly within her paraspinal musculature, an enlarged T-5 rib on the right, and a flow void within the spinal canal at the T-5 level compressing and displacing the cord to the left (Figs. 1 and 2). The T2 signal change was present above and below this flow void, which suggested venous congestion (Fig. 3). Numerous flow voids were also noted within the complex, fatty mass posterior to her thoracic spine (Fig. 4).

She was transferred to our institution for further evaluation and treatment. She was given steroids, and proprioception and strength improved in her right foot. Spinal angiography was performed, which revealed a highly vascular metameric-like lesion with an epicenter at the T-5 level with numerous abnormal vascular feeders coming off the T-4, T-6, and T-7 segmentals bilaterally (Fig. 5).

Operation. The patient was positioned prone on the operating room table, and through a midline incision an extended bilateral dissection was carried out, extended on the right to allow for exposure of the large paraspinal mass. This mass was noted to extend from T-4 to about T-10, and it wrapped laterally around the patient’s chest wall. Full dissection was carried out around the entire lesion, which was noted to be very hemorrhagic. Bilateral subperiosteal dissections were performed to allow for full exposure of the posterior elements bilaterally from T-4 to approximately T-10 with extended exposure on the right.

There were multiple, high-flow epidural fistulous connections, with vascular channels traversing mainly the T-5 and T-6 VBs and draining into the azygous and hemiazygous systems. Of note, the patient’s anterior spinal artery was fed by the T-6 segmental on the left side.

The lesion was partially embolized, and she exhibited gradual improvement neurologically. Steroids were continued. After 2 weeks of rehabilitation, she was able to take individual steps on her own. The patient’s progress plateaued, and repeat embolization and definitive resection were undertaken.

Fig. 1. Sagittal T1-weighted MR image showing large paraspinal mass mainly posterior to the T5–9 VBs that contains fat and prominent flow voids. Adjacent to the spinal cord, there is another flow void.

Fig. 2. Noncontrast axial CT scan showing the T-6 VB with enlarged rib head and transverse process on the right, with bony compression of the spinal cord, displacing the cord to the left.
Metameric thoracic lesion

exposing the chest wall. After the lesion was dissected free, the paraspinal mass was then removed first from the chest wall and then back toward the spinal column. The lesion was then truncated at the level of the spinal column and sent to pathology for evaluation.

Attention was then turned to performing a decompressive laminectomy from T-3 to T-7 and costotransversectomy at T-5. This was done in the standard fashion using a high-speed drill and rongeurs. The bone was noted to be extremely hemorrhagic with several venous varices, which were controlled using bone wax. Additional bleeding was noted from engorged epidural vessels, which was controlled with bipolar electrocautery. Pediculectomies were then performed at T-4, T-5, and T-6 to allow for full exposure of the epidural component of the malformation. This was then removed using a combination of bipolar cautery, microscissors, and vascular hemoclips. The left T-6 radicular artery was preserved.

Despite 2 prior aggressive embolization procedures, this lesion was still extremely vascular, and the patient required an intraoperative transfusion of 16 U of packed cells in addition to fresh-frozen plasma and platelets. After our resection, the wound was closed in a multilayered fashion by plastic surgery with mobilization of a trapezius flap to help close the dead space that remained after removal of this massive lesion (Fig. 6).

Postoperative Course. Over the course of her 2-week postoperative hospital stay, the patient improved neurologically. She had full strength in her lower extremities, her joint position sense returned to normal, and she was ambulatory. Postoperative CT scanning was done, which showed good decompression of her spinal canal and no obvious remaining lesion (Fig. 7). A postoperative spinal angiogram showed only a small area of arteriovenous shunting anterior to the VB at T-4, but no epidural fistula or residual AVM was noted.

After 8 months, the patient remains slightly hyperreflexic in her lower extremities, but she has full strength in all muscle groups with intact proprioception. Magnetic resonance imaging performed at that time did not show any definitive evidence of a residual AVM, but it did show resolution of the previously seen T2 signal change in the cord (Fig. 8).

Pathological Examination. The gross specimen consisted of 2 irregular portions of yellow, lobulated adipose tissue; the larger piece measured 31 × 22 × 6.5 cm and the smaller piece measured 14 × 10 × 2 cm (Fig. 6 upper). The cross-sections of the specimen showed yellow nodular surfaces with distinct dilated vascular channels (Fig. 6 lower). Microscopic examination revealed a hamartomatous lesion that was predominantly composed of mature adipose tissue with embedded complex vascular malformations, atrophic skeletal muscle tissue, and nerve twigs (Fig. 9). Some of the vascular malformation lesions consisted of complex thick-walled interconnected vascular channels. Special stains for elastic and trichrome demonstrated presence of arterial and venous type vessels. The lumens of many other cavernous appearing ectatic vascu-
lar lesions contained complex interconnected capillary-like vascular lumens lacking internal elastic lamina.

**Discussion**

Metameric lesions tend to be complex, and their treatment will optimally involve a multidisciplinary approach. Preoperative imaging should include MR imaging and CT scanning to assess bony involvement, and spinal angiography with extensive preoperative embolization. In this case, even after 2 thorough embolizations, blood loss was significant, emphasizing the critical role embolization plays in the treatment of these lesions. In the thoracic spine, as in this case, angiography also proved invaluable to identify the location and arterial supply for the anterior spinal artery; proper identification minimized the chance of spinal cord infarct both during embolization as well as during resection.

Although these lesions are benign in pathology, patients may develop symptoms as a result of direct spinal cord compression and/or venous hypertension. In this case, our patient was myelopathic likely from long-term bone compression at the level of the lesion, as well as from a more acute change in the vascular behavior of the lesion, leading to venous hypertension, the extensive T2 signal change seen on MR imaging, and her history of progressive gait difficulty over a 2-week period.

Published reports have stated that metameric spinal lesions require mechanical stabilization after resection. We did not recommend fusion in this case. Our patient had obvious transosseous vascular channels, but her bone quality appeared solid on preoperative and intraoperative CT scanning. Additionally, the lesion was in the thoracic spine, and because the rib cage provided enough stability a stabilization procedure was unnecessary. Also, because we wanted to obtain immediate postoperative as well as
delayed spinal angiograms and serial MR images, we did not want to have our imaging degraded by metal artifact from any instrumentation, particularly for a lesion with unknown long-term behavior. The patient’s 8-month follow-up MR imaging study showed the development of a mild kyphotic deformity (Fig. 8), but she is asymptomatic from this with no back pain or pulmonary complaints. We plan to observe her clinically and radiographically; if the lesion should return and/or if she should develop a significant kyphotic deformity, our plan is to undertake another resection and/or correct the deformity.

**Conclusions**

Metameric spinal lesions are rare, complex entities that are optimally approached in a multidisciplinary fashion. Left untreated, these lesions may lead to progressive myelopathy due to direct cord compression as well as from congestion or an arterial steal phenomenon. Excellent preoperative embolization is the key to successful resection and good patient outcome.
Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References

1. Adegboyega PA, Qiu S: Hemangioma versus vascular malformation: presence of nerve bundle is a diagnostic clue for vascular malformation. Arch Pathol Lab Med 129:772–775, 2005