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Delayed en bloc excision of L3 for metastatic sacrococcygeal teratoma on a 1-year-old boy - a case report.

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Abstract

Case: A 1-year-old boy was presented with cauda equina syndrome and progressive loss of motor function in lower limbs. MRI and CT scans revealed a sacrococcygeal teratoma with metastases para-aortically and in L3 producing compression into the epidural space. Despite metastases and a progressive cauda equina, neoadjuvant treatment was given to achieve cytoreduction for neurological recovery and facilitate curative treatment

Conclusion: Malignant sacrococcygeal teratoma with intraspinal infiltration or metastasis requires surgical and adjuvant treatment. If primary neurologic function-sparing surgery makes curative treatment impossible, adequate and quick decompression of neural structures with similar results can be achieved by preoperative chemotherapy, avoiding the local spill of malignant cells.

Yolk sac tumors (YST) are germ cell tumors most commonly arising from the gonads, but extragonadal YST also occur1. YST also occur in combination with teratoma[1]. Mature teratoma is a benign tumor, whereas YST are mostly malignant. Because YST can occur as small malignant foci, needle biopsy can be negative. In such cases, alphafetoprotein (AFP) aids in differential diagnosis because elevated levels of AFP suggest malignancy.

Sacrococcygeal teratoma (SCT) is a commonly benign neoplasm adjacent to the sacrum and coccyx. SCTs are categorized in 4 types depending on intrapelvic and extrapelvic distribution (Altman classification). Type I and II SCTs are easily detected and palpable and most often diagnosed immediately after birth. The risk of malignancy rises quickly after the age of 2 months (7% to 10% risk at less than 2 months and 80% to 90% risk at older than 2 months)[2]. Malignant SCTs with yolk sac components generally respond well to chemotherapy and express high levels of AFP, which aids in diagnosis and is useful during follow-up[1-4]. Significant risk factors for metastasis are Altman type III-IV and older age at time of diagnosis. Both mature, immature, and malignant teratoma can be metastatic. Typical sites of metastasis include the liver, lungs, lymph nodes, and spine[5]. The risk of recurrence increases with incomplete resection and immature or malignant histology.

The patient and his parents were informed that data concerning the case would be submitted for publication, and they provided consent.

Case Report

1-year-old previously healthy boy started to show signs of decline in motor functions and overall distress. He had recently learned to walk against support, but over a 2-week period, he regressed to crawling and then proceeded to move less and less. The boy was crying more than usual and presented with abdominal distension. He was referred to the emergency department and was diagnosed with urinary retention of 280 mL (median bladder capacity for a 1-year-old child is 60 mL). Catheterization led to clearly diminished distress. Over the next 2 days, he presented with rapid loss of motor function in legs and diminished tendon reflexes accompanied by sphincter paralysis. A full-body magnetic resonance imaging (MRI) was performed to investigate the etiology of cauda equina syndrome.

The MRI showed a tumor (3.4 · 4.2 · 5.4 cm) in the pelvis located between rectum and sacrum compressing both the rectum and urethra. Other tumor masses were found in the third lumbar vertebra, in the epidural space, and in a para-aortic mass. The epidural mass in the spinal column was pressing the cauda equina at L2 to L3 level (Fig 1). A whole-body computed tomography (CT) scan showed several metastases in the lungs. Initially, the tumor was suspected to be metastatic neuroblastoma.

Curative treatment plan was discussed between pediatric oncologists, pediatric surgeons, pediatric orthopaedic surgeons and neurosurgeons, and the parents of the patient. Although treatment of cauda equina syndrome typically involves emergency surgery, it was considered a great risk, allowing for further spread of cancer by local spill of malignant cells and jeopardizing curative treatment. It was conceded that preoperative chemotherapy and postponed surgery was an option

because the onset of symptoms had been relatively slow, and the nerves of the cauda typically are more resilient to pressure than the spinal cord.

Steroid treatment was initiated immediately, a CT-guided biopsy was taken from the lesion in L3 corpus and an ultrasoundguided biopsy from the pelvic tumor mass. The frozen section biopsies ruled out neuroblastoma, and treatment with Linesprotocol (etoposide 1 carboplatin) was initiated targeting both sarcoma, neural malignancies, and germ cell tumors. Histological analysis showed high mitotic activity and severe atypia. Multiple histologic patterns, including solid, glandular, microcystic, and papillary patterns, were represented. Immunohistochemical staining was positive for AFP, CD117, and pancytokeratin. SMA, HCG, CD30, Oct4, podoplanin, and S-100 were negative. Further, Ki-67 showed a proliferation index of 90%. The levels of AFP were >70,000. Histopathologic results were the same in both biopsies, confirming metastatic stage IV malignant SCT with yolk sac component (Fig 2).

The patient showed a moderate increase in motor functions on the first day after initiation of treatment with steroids and chemotherapy. The chemotherapy plan was modified to meet the guidelines of YST treatment: 6 rounds of carboplatin, etoposide, and bleomycin (JEB treatment). After 2 months of JEB treatment, a follow-up MRI scan showed that the sacral tumor had shrunk significantly. The boy started crawling again. After 4 months of chemotherapy, the follow-up MRI and CT scans showed significant tumor reduction in the pelvis, 1.6 · 1.0 · 2.6 cm (initially 3.4 · 4.2 · 5.4), and the para-aortic, pulmonary, intraspinal, and vertebral metastases were not detectable. The L3 vertebrae had collapsed after tumor lysis. AFP levels were normal, indicating a good response to chemotherapy.

After 20 weeks (6 cycles) of chemotherapy, pelvic surgery was performed. Laparoscopic dissection of presacral space was performed, followed by open surgery from a posterior sagittal approach in the same session. The coccyx was severed from the sacrum, and the presacral tumor was then dissected from the rectum and Waldeyer's fascia. The tumor was macroscopically yellow and necrotic. After removal of the tumor, the anterior aspect of the sacrum was shaved superficially to determine tumor margins.

At 24 weeks after onset of chemotherapy, en bloc spondylectomy was performed using a combined approach. The procedure was started with posterior exposure, L3 laminectomy, clearing epidural space, and L3 nerve roots were protected. Pedicle screws were implanted at L2 and L4 using 3.5-mm posterior cervical instrumentation with short 3.5-mm Titanium Alloy rods. Left-sided thoracoabdominal approach was performed including L2/3 and L3/4 complete discectomies, L3 en bloc spondylectomy (Fig. 3), and reconstruction using allograft (humeral diaphysis). A thoracolumbosacral brace was used 6 months postoperatively. Histopathological analysis of the primary tumor and L3 metastasis both confirmed 100% tumor necrosis with no malignant cells and clean margins.

Thirty months from diagnosis, 24 months postoperatively, the boy is now 3.5 years old, walking normally for his age, and has normal patellar and Achilles reflexes. Bladder function is still impaired, and catheterization a few times per day is necessary, but some spontaneous urination does occur.

AFP levels remain low, and follow-up imaging (latest MRI performed at 23 months postoperatively) has not detected any signs of recurrence.

Discussion

is the most common germ cell tumor in neonates[4,6] and, overall, one of the most common pelvic neoplasms in neonates and children[2]. Considering the occurrence at 1: 35,000 to 40 000 live births[4,5,6] it is still rare. Altman type IV SCTs (with only intrapelvic tumor) only comprise about 15 to 28%5,6 of all SCTs, and only about 8% to 15% of SCTs have intraspinal extension or metastases[3,4]. The rate of malignant SCT is much higher in patients older than 2 months[2], suggesting that early diagnosis and removal are key in preventing malignancy. Stage IV Altman tumors are considerably more common in older age groups[5,7], which could be a result of difficult diagnosis because of nonpalpable masses and consequent malignant transformation of previously benign tumors.

In our patient, the first symptoms of the tumor were those of lumbar nerve and cauda equina compression, and compression of the urethra and rectum. The symptoms progressed considerably over 2 weeks, suggesting substantial tumor growth. After onset of chemotherapy, the symptoms also regressed rapidly, signalling tumor shrinkage. It is essential to start chemotherapy without delay when an Altman type IV tumor is diagnosed.

In patients with epidural tumor components, spinal tumor surgery cannot, by definition, be radical. Most malignant SCTs and yolk sac tumors respond well to chemotherapy[1,3,4], and even with intraspinal extension, tumor response can be so remarkable that exploration of the spinal canal is

rendered unnecessary[3]. In our case, however, a separate metastasis had led to collapsed vertebrae during chemotherapy because of tumor lysis of intravertebralmass. Excision of L3 and fusion was necessary to achieve optimal local tumor control, a stable spine, and return of motor function. This patient represents the youngest patient in literature undergoing en bloc excision in the thoracolumbar spine.

Despite delayed surgical intervention, full neurological recovery in lower extremity motor functions was achieved, in combination with relapse-free survival for 2 years postoperative follow-up. Impaired bladder function is common in children treated for SCT[4,6,8] and thus is not necessarily considered to be caused by delayed surgical intervention.

Conclusion

When treating malignant SCT, a multidisciplinary team is vital to avoid diagnostic errors and optimize treatment. Cauda equina symptoms usually demand emergency decompression to preserve sensorimotor functions in lower limbs and avoid incontinence and urinary retention. It is important to differentiate cauda equina symptoms with slow, progressive onset that are caused by a tumor, e.g., SCT, that can rapidly respond to chemotherapy and steroids thus delaying surgery and minimizing its inherent risks of local and systemic tumor progression[9].

References

1. Kattuoa MI, Dunton CJ. Yolc Sac Tumours. StatPearls Publishing LLC; 2023.

2. Groff DB. Pelvic neoplasms in children. J Surg Oncol. 2001;77(1):65-71.

3. Garg R, Agarwala S, Bakhshi S, Srinivas M, Bajpai M, Gupta DK, Bhatnagar V. Sacrococcygeal malignant germ cell tumor (SC-MGCT) with intraspinal extension. J Pediatr Surg. 2014;49(7):1113-5.

4. Seilern Und Aspang J, Burnand KM, Ong H, Cross K, Thompson D, Giuliani S. Sacrococcygeal teratoma with intraspinal extension: a case series and review of literature. J Pediatr Surg. 2020;55(10):2022-5.

5. Derikx JPM, De Backer A, van de Schoot L, Aronson DC, de Langen ZJ, van den Hoonaard TL, Bax NMA, van der Staak F, van Heurn LWE. Factors associated with recurrence and metastasis in sacrococcygeal teratoma. Br J Surg. 2006;93(12): 1543-8.

6. Salim A, Raitio A, Losty PD. Long-term functional outcomes of sacrococcygeal teratoma: a systematic review of published studies exploring ‘real world’ outcomes. Eur J Surg Oncol. 2023;49(1):16-20.

7. Rescorla FJ. Pediatric germ cell tumors. Semin Pediatr Surg. 2012;21(1):51-60.

8. Rescorla FJ, Sawin RS, Coran AG, Dillon PW, Azizkhan RG. Long-term outcome for infants and children with sacrococcygeal teratoma: a report from the Children’s Cancer Group. J Pediatr Surg. 1998;33(2):171-6.

9. Mirzaei L, Kaal SE, Schreuder HW, Bartels RH. The neurological compromised

spine due to ewing sarcoma. What first: surgery or chemotherapy?

Therapy, survival, and neurological outcome of 15 cases with primary Ewing sarcoma of the vertebral column. Neurosurgery. 2015;77(5):718-25; discussion 724-5.

Figures

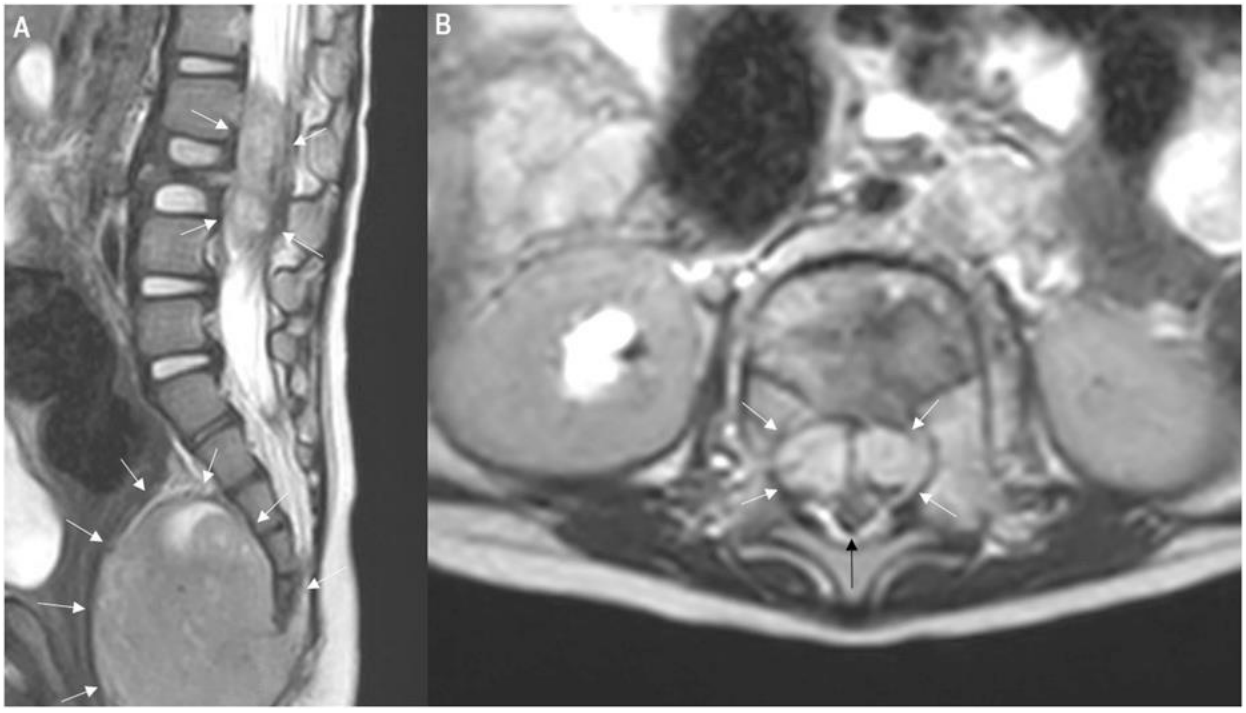

Figure 1. Magnetic resonance T2 images of the spine: Sagittal view (Fig. 1-A) showing a large presacral tumor, epidural tumor mass on L2 to L4 level, and collapsed
L3 vertebra (both marked with white arrows). Axial view (Fig. 1-B) on L2 to L3 level showing tumor mass in the spinal canal (white arrows) and cauda equine
compressed posteriorly (black arrow).

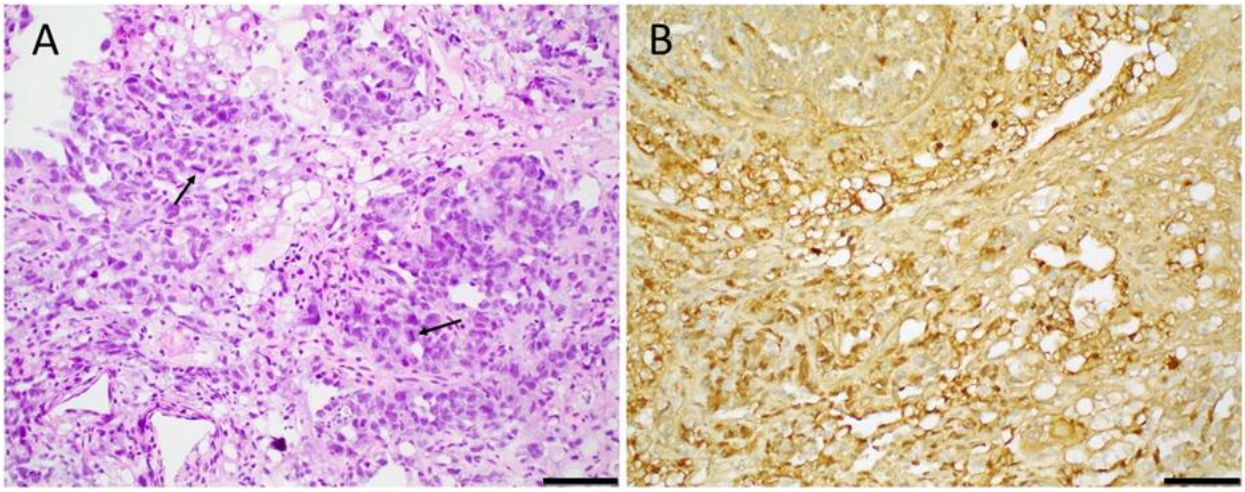

Figure 2. Histology of malignant yolk sac tumor component. Fig. 2-A Hematoxylin and eosin staining. Cytologic atypia is prominent, and mitoses are present (arrows).

Occasional tumor cells contain lipids. Fig. 2-B AFP immunohistochemical staining is diffusely positive in tumor cells. This finding is highly specific to yolk sac

tumors. Scale bars: 50 mm. AFP = alpha-fetoprotein.

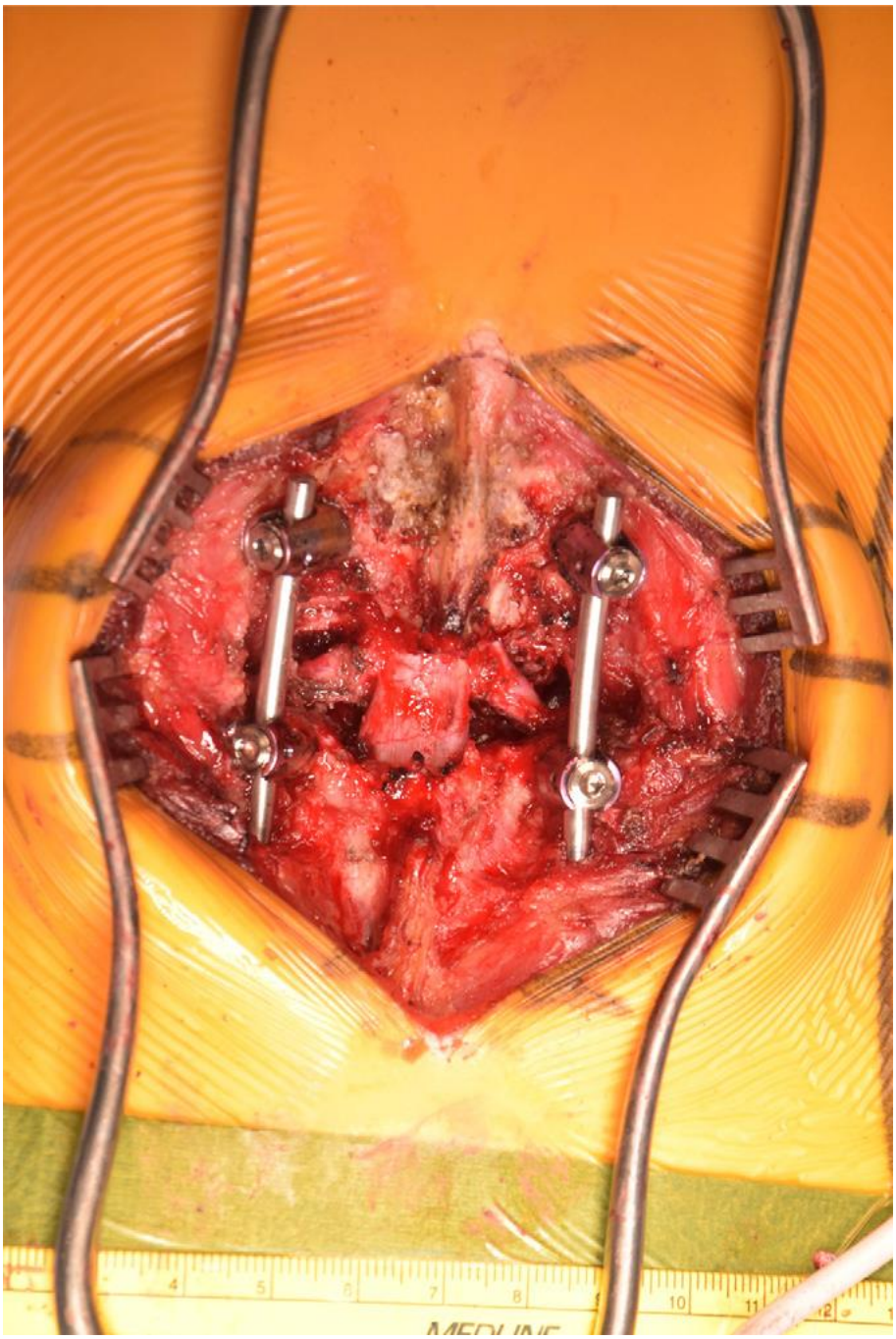

Figure 3. En bloc excision of L3. Posterior approach.