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Aggressive hemangioma of the spine in a pregnant female: a case report and literature review

Gebe bir kadında omurganın agresif hemanjiomu: Olgu sunumu ve literatür taraması

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ABSTRACT

Type and timing of treatment for symptomatic hemangiomas in pregnant females are challenging due to fetus survival and conflicts in neurological recovery. In this article, we report a 40-year-old female patient at pregnancy week 23 with a complicated hemangioma at T_1 level. Physical examination revealed an incomplete spastic paraplegia. Patient did not accept any surgery due to child's death risk. Patient was started corticoid treatment and no more weight bearing was allowed. At the 28th week of pregnancy, the patient underwent cesarean section immediately followed by selective arterial embolization, decompression, fixation, and radiotherapy. At two-year follow-up, the patient was pain free, without any signs of local recurrence and with complete neurological recovery. A multidisciplinary approach is mandatory to save the life of the fetus without damaging the spinal cord functions of the mother.

Keywords: Hemangioma; paraplegia; pregnancy; spine.

ÖZ

Gebe kadınlarda semptomatik hemanjiomların tedavi tipi ve zamanlaması fetüsün yaşaması ve nörolojik düzelmedeki tartışmalar nedeniyle zordur. Bu yazıda, T_1 seviyesinde komplike olmuş hemanjioması ve gebeliğinin 23. haftasında olan 40 yaşında bir kadın hasta sunuldu. Fizik muayenede inkomplet spastik parapleji görüldü. Hasta, çocuğun ölüm riski nedeniyle hiçbir ameliyatı kabul etmedi. Hastada kortikoid tedavisine başlandı ve daha fazla ağırlık taşımasına izin verilmedi. Gebeliğin 28. haftasında hastanın sezaryen ile doğumu gerçekleştirildi; takiben selektif arteriyel embolizasyon, dekompresyon, fiksasyon ve radyoterapi uygulandı. İki yıllık takibinde hasta herhangi bir lokal nüks olmaksızın ve tam nörolojik iyileşme ile ağrısızdı. Annenin spinal kord fonksiyonlarını bozmadan çocuğun hayatını kurtarmak için multidisipliner bir yaklaşım şarttır.

Anahtar sözcükler: Hemanjiom; parapleji; gebelik; omurga.

Vertebral hemangiomas can become symptomatic in pregnancy. Review of the literature for vertebral body hemangiomas in pregnancy revealed 23 cases in 21 case reports leading to neurological deterioration (Table I).[1-21] Time from onset of symptoms to intervention had a wide variation, from two days to six months. Emergency care can be easily undertaken in these patients; however, some challenges emerge when the patient is at the second term. In this article, we report a paradigmatic case of a pregnant female with multiple challenges.[22]

CASE REPORT

A 40-year-old pregnant female patient was referred to the emergency room at the 23th week of gestation with cervicothoracic pain and gradually increasing weakness at lower limbs that started three weeks before. The patient was unable to walk and there was a sensory loss up to the shoulders. The motor examination revealed spastic incomplete paraplegia with proximally 2/5 and distally 4/5 motor strength in both legs. Babinski test was bilaterally positive,

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TABLE I

Systematic review of the literature for complicated vertebral body hemangiomas in pregnancy

Authors	Gestation	Level	Duration of symptoms	Recovery
Guthkelch[21]	34	T_6	1 month	Death
Askenasy and Behmoaram[20]	34	T_{10}	15 days	Complete
Fields and Jones[19]	28	T_{10}	3 months	Complete
Newman[18]	32	L_3	8 months	Complete
Newman[18]	36	T_4	1 month	Complete
Newman[18]	32	T_{4-5}	3 months	Death
Nelson[17]	28	T_{2-4}	1 month	Partial
Esparza et al.[16]	24	T_{5-7}	2 months	Complete
Faria et al.[15]	32	T_4	6 months	Complete
Lavi et al.[14]	28	T_{4-6}	1 month	Partial
Schwartz et al.[13]	30	T_5	1 month	Complete
Liu and Yang[12]	20	T_4	1 month	Complete
Redekop and Del Maestro[11]	32	T_{12}	4 months	Partial
Tekkök et al.[2]	Po	T_5	40 days	Complete
Castel et al.[5]	28	T_8	few days	Partial
Chi et al.[4]	24	C_7	25 days	Partial
Inamasu et al.[6]	33	L_2	10 days	Complete
Yüksel et al.[9]	28	T_9	2 months	Complete
Vijay et al.[7]	26	T_{11}	8 days	Complete
Kiroglu et al.[8]	36	T_4	few days	Complete
Schwartz et al.[3]	Po	T_{11}	2 days	Complete
Shinozaki et al.[10]	28	T_2	few days	Complete
Blecher et al.[27]	37	L_4	several weeks	Complete
Present case	23	T_1	8 weeks	Complete

Duration of symptoms: Time from onset of symptoms and operative intervention; Po: Postpartum.

there was mild clonus on both legs and both knee jerk and Achilles reflexes were bilaterally exaggerated. The examination of the upper extremity exhibited 4/5 motor strength at the left interosseous muscles. Computed tomography (Figure 1) with three-dimensional reconstruction (Figure 2) and magnetic resonance imaging (Figure 3) were obtained. Computed tomography showed honeycomb pattern involving entire T_1 vertebra strongly suggestive for hemangioma. A written informed consent was obtained from the patient.

On admission, emergency surgical decompression was proposed to the patient, immediately after interruption of the pregnancy. The patient refused interruption of pregnancy and any other treatment possibly creating high risk for the fetus survival. She was alerted of the risk of worsening of neurological conditions and irreversible paraplegia.

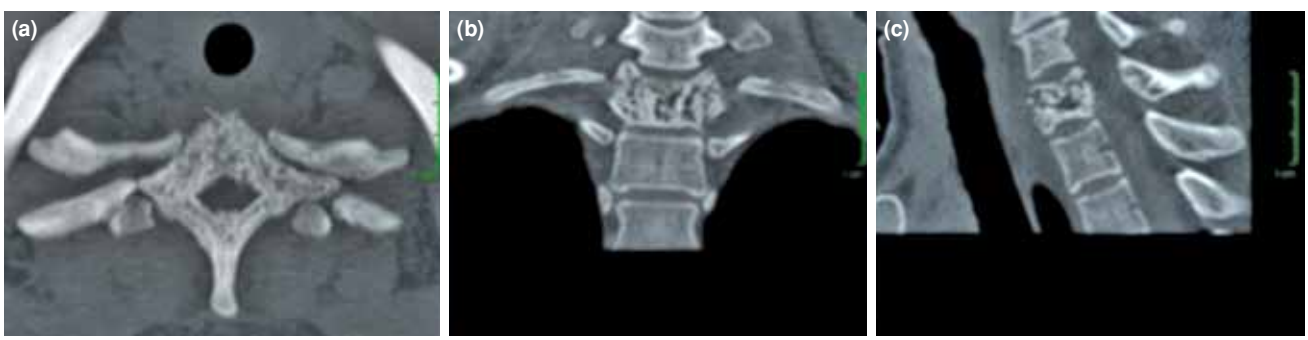

Figure 1. (a) Axial, **(b)** coronal, and **(c)** sagittal computed tomography sections of T_1 vertebra show classical appearance of a hemangioma with vertical striations and honeycomb pattern involving both corpus and posterior neural arch of T_1.

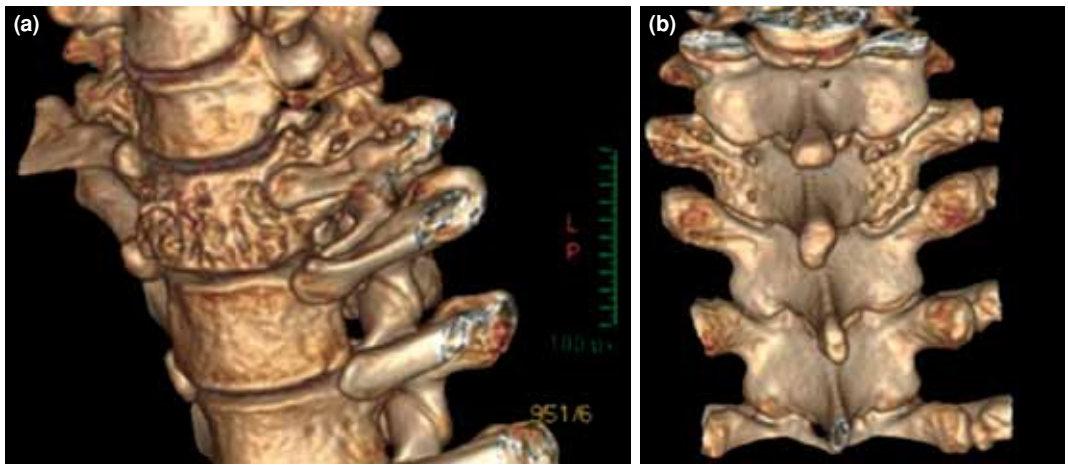

Figure 2. (a) The anterolateral and **(b)** posterior views of three-dimension computed tomography of cervicothoracic junction demonstrates involvement of entire T_1 vertebra.

The decision making process also involved a gynecologist, a radiotherapist, and an interventional radiologist. The final decision was to keep the patient lying in bed under corticoid treatment until the fetus maturity detected by ultrasound imaging and functional exams could allow performing a caesarian operation.

The patient accepted and was started 4 mg intravenous betamethasone per day, weight bearing was not allowed, but motor rehabilitation program included active muscular exercises. At the 28th week of pregnancy, angiography and embolization was performed, achieving 85% devascularization. On the following day, she was first submitted to a cesarean section under general anesthesia, and then cord decompression by laminectomy and transpedicular resection of the tumor followed by C_4-T_3 stabilization and fusion (Figure 4). The newborn was a male with excellent life parameters. The patient gradually started standing and walking exercises. Neurological status gradually improved. Six weeks after surgery, she received external beam conventional radiation therapy. At six months, she was ambulatory without assistance. Histopathological diagnosis had confirmed cavernous hemangioma.

At two-year follow-up, there is no evidence of local progression of the hemangioma, the neurological function is normal, and patient is fully ambulant and she returned back to work and social life. The child is fully normal.

Figure 3. (a) Axial T_2-weighted noncontrast-enhanced, **(b)** sagittal T_2-weighted noncontrast-enhanced, and **(c)** sagittal T_1-weighted magnetic resonance images of patient obtained preoperatively which were heterogeneously hyperintense demonstrate severe compression of spinal cord by epidural extension of tumor.

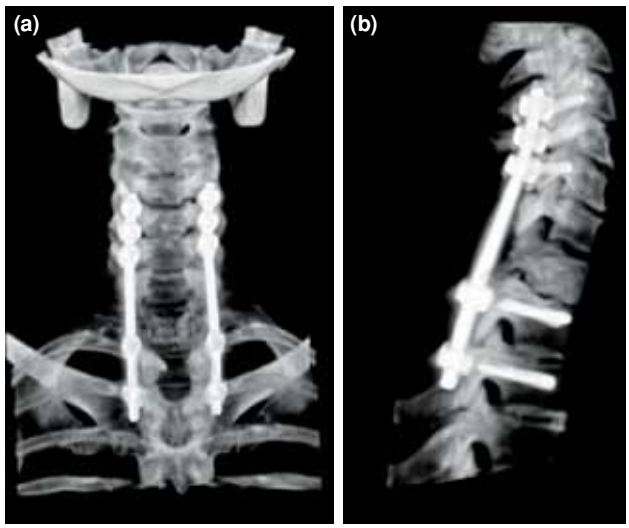

Figure 4. (a) Coronal and **(b)** sagittal postoperative computed tomography images show well decompressed and stabilized spine without any recurrence at two-year follow-up.

DISCUSSION

Vertebral hemangiomas are usually discovered incidentally; 10% to 12% of are reported to occur in a thoracic vertebra.[23] The epidemiology, diagnostic characteristics, and management of these benign spinal neoplasms have been extensively discussed in the literature.[24-27] The possibility of hemangiomas increasing in size, compressing the cord and reducing the vertebral body resistance during pregnancy or puberty is well known and is related to altered progesterone and estrogen levels and/or obstruction of paravertebral veins draining into inferior vena cava by gravid uterus.[28,29] When hemangioma becomes symptomatic in a pregnant female, decisions related to timing and type of treatment are challenging due to the conflicting interests of neurological recovery (and treatment of pathologic or impending fracture) and fetus survival.

The case reported herein concerns a pregnant female complaining of pain and severe neurological problems at the 23rd week. Patient refused to undergo emergency decompression which may expose the fetus to life risk mostly due to possible profuse bleeding while resection of the tumor. Cesarean operation at that time would as well end with the death of the fetus. Even if laminectomy could have been performed and pregnancy continued, the hemangioma would have been growing due to the continuity of hormonal activity. Radiotherapy alone was contraindicated for its teratogenic effect. The decision to delay decompressive surgery under corticoid treatment was also favored by the demonstrated association between exposures of low doses of betamethasone and accelerated fetal lung maturation.[30] As soon as maturation was acceptably defined, cesarean operation and cord decompression were performed on the same day. Selective embolization the day before and radiotherapy after six weeks completed the treatment.

In conclusion, dealing with a complicated hemangioma in a pregnant female encompasses several issues: the risk of permanent paraplegia compared to the risk against the life of the fetus, increased risk of intraoperative profuse bleeding or radiation exposure of the fetus from CT scan during embolization or radiotherapy. A multidisciplinary approach included the spine surgeons discussing the case with a gynecologist, a pediatrician, an interventional radiologist, a radiotherapist, and obviously the patient and her family. The review of the literature for complicated hemangiomas in pregnancy revealed that vast majority of the cases had symptoms for several weeks or months. Emergency surgery can be delayed while keeping the patient under strict neurological observation, till the maturity of the fetus.

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