Spinal angiolipoma

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Citation

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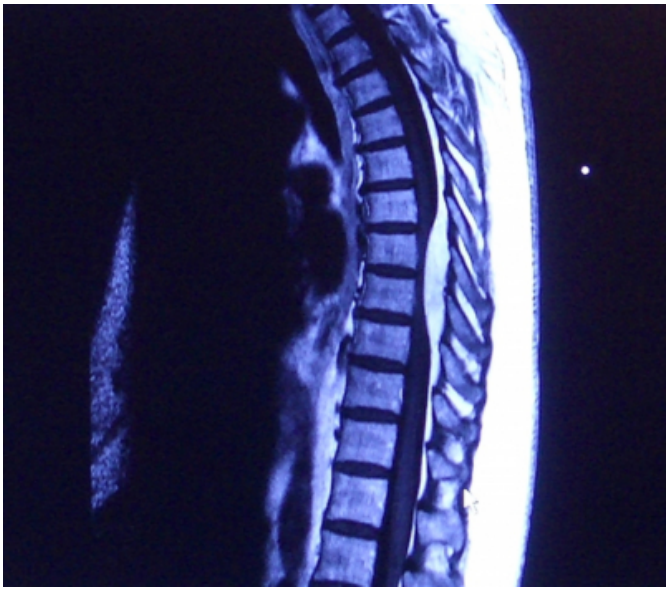
Abstract

Background; Spinal angiolipomas are rare benign tumors composed of mature lipocytes admixed with abnormal blood vessels. Spinal angiolipomas approximately accounts for 0.14 % to 1.2 % of all spinal axis tumors, about 2-3% of extradural spinal tumors. Extradural tumors usually predominate. It commonly presents with local spinal pain, spastic or peripheral paraparesis of the lower extremities, paraesthesia and impaired sensitivity of the cord or root type.Methods;A case of spinal angiolipoma in a 58 -year-old healthy male patient who presented with chronic epigastric discomfort for 3 years and sudden onset of acute myelopathy. MRI confirmed a space occupying lesion identified at thoracic 7, 8 and 9 vertebrae levels. It was posterior in location with spindle shape measuring 6.5 cm in length. A laminectomy was performed with gross total resection of the mass. Pathology was consistent with angiolipoma.Results; The patients neurological symptoms improved. Follow-up scan did not show any tumor and clinically no neurological deficit.Conclusion; MRI is investigation of choice. The main stay of treatment is complete surgical extirpation. Radiation is not necessary even if patient has residual tumor as prognosis is good regardless.

CASE REPORT

A 58 years old healthy male had chronic pain in upper abdomen radiating to back for past three years. He had no other systemic compliant or any other medical co morbidity. He was investigated for this pain and was diagnosed as irritable bowel syndrome. He used to take analgesics off and on and was quite frustrated with this nagging pain which was affecting his quality of life. However he was still active doing his job and attending to his daily chores. He suddenly developed bilateral leg weakness which got worst over 24 hours and reported to local hospital emergency room. In ER his symptoms aggravated and he developed urinary retention and loss of rectal continence. Tracing his history back, he had this epigastric nagging pain since last 36 months radiating to upper thoracic spine. The epicenter as per patient was in gastric area. No back injury or neurological symptoms in past 36 months. Patient did some heavy lifting before he suffered symptoms and it may have increased resistance of venous return during the act. Physical examination revealed paraparesis .The sensory impairment was below Thoracic 10 level with no sacral sparing. Deep tendon reflex of lower legs had increased. No distinct family history or hereditary disease which might put him at risk. No dysraphism was seen. Rest of the systemic examination was essentially normal. MRI of the spine (figure 1) showed space occupying lesion identified at thoracic 7, 8 and 9 vertebrae levels. It was posterior in location with spindle shape measuring 6.5 cm in length and about 10mm at its maximum width.T1 and T2 sequences and post contrast with fat suppression suggested it to be a meningioma. Rest of the spine was normal.

Figure 1



The patient underwent T 7-9 laminectomy with total removal of epidural mass. Post operative period was uneventful.

Pathology report revealed sections with benign adipose tissue with a prominent vascular network .Vessels were very congested and predominantly capillaries with some fibrin thrombi. The cellular variant included angiomatous tissue,

spindle cells abundant in cellular areas and associated with mild pleomorphism. The picture was consistent with angiolipoma (figure 2 and 3). No evidence of malignancy. Three months later patient had full neurological recovery. Epigastric pain had completely subsided. Follow up MRI (figure 4) showed post operative changes, no residual tumor. Spinal canal well maintained.

Figure 2

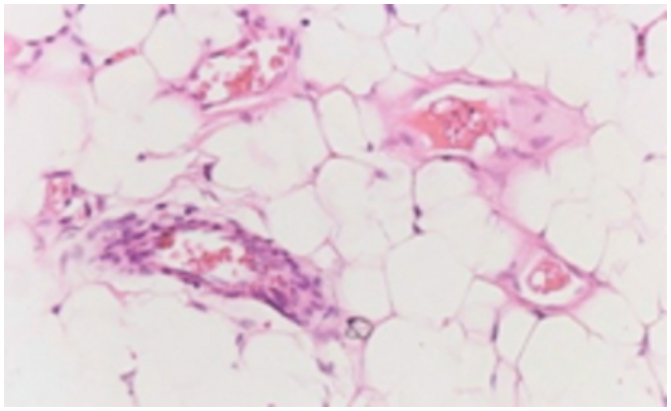


Figure 3

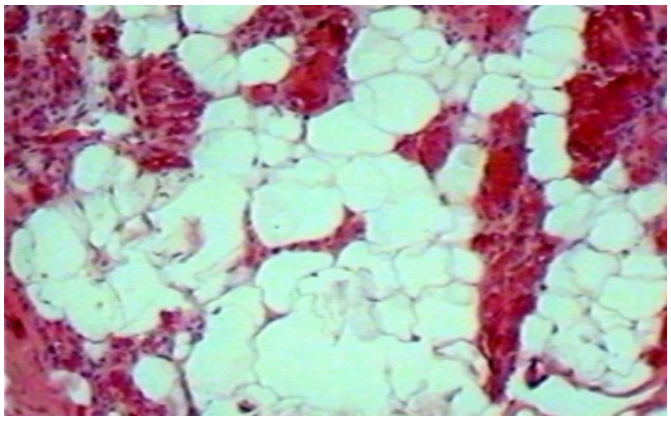
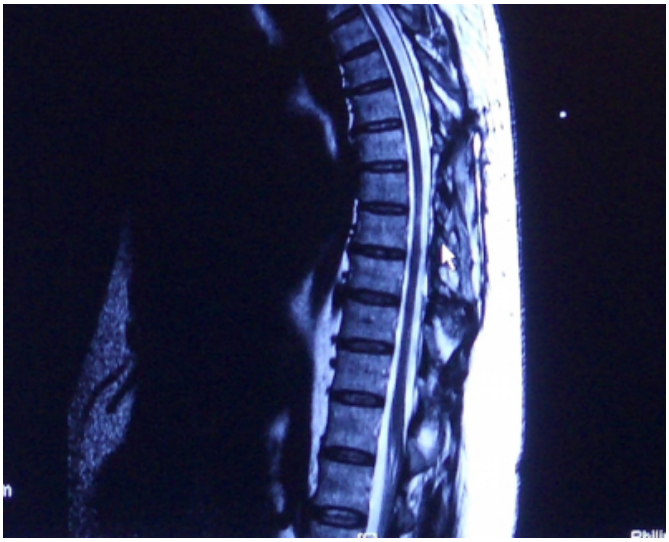


Figure 4



The patient has returned back to work and is fully active at 12 month follow up.

DISCUSSION

Approximately 80 cases of spinal angiolipomas have been described in the English medical literature since 1996. Dr Liebscher in 1901 was the first clinician to describe angiolipoma of the spine with clarity[1]. Later in 1960 Howard and Helwig established angiolipoma as distinct entity[2]. Spinal angiolipomas are rare benign lesions composed of mature lipocytes cells admixed with vasculature element. Spinal epidural angiolipomas predominantly affect women and middle aged males[3]. A slow progressive clinical course is seen and rarely a fluctuating course In pregnant females have been documented in the literature. The symptoms in pregnant females are probably exacerbated by weight gain, suggesting that vascular engorgement, presence of obesity influencing the evolution. Their preponderance in older, peri or postmenopausal women and clinical exacerbation in pregnant women support a role of hormonal influence[4]. Based on studies by Lin and Lin, angiolipomas are subdivided into two histological types: infiltrating and non infiltrating. Infiltrating angiolipoma is characteristically not encapsulated, and they infiltrate into surrounding tissue. Their clinical behavior is similar to that of hemangioma. Prognosis is good in both types if removed completely[5]. Infiltrating angiolipomas are usually diagnosed in older patients. The vast majority occur in the lower extremities or in the paraspinal region, which can lead to muscular pain and neural deficits[6]. Majority of angiolipoma are epidural based but there are case reports of spinal intramedullary angiolipoma. Non infiltrating or circumscribed angiolipomas

are encapsulated lesions limited to the subcutaneous compartment. Their size almost never exceeds 4 cm. These lesions are more common in young people, and they are equally distributed between the sexes[7]. Trabulo et al reported eight out of 11 cases of infiltrating angiolipoma were partially removed and neurological improvement was seen in all of them. None of them required further surgery[8].

Regarding investigations myelogram is abnormal in approximately 40% and shows a complete block in 63% of the patients. A CT appearance of the spinal angiolipoma is that of a mass that is typically hypo dense relative to the spinal cord and has variable degree of enhancement. Rarely may it show calcification. Vertebral bodies infiltrated by disease show little or no contrast enhancement, which is a reliable indicator to distinguish them from hemangioma[9]. Contrast may show enhancement of the epidural angiolipoma. The MRI demonstrates a fatty mass with an abundant vascular component suggesting the diagnosis. The mass is usually hyper intense on T1 than T2-weighted signals, but can also be heterogeneous or mixed intensity. .It may be isointense with epidural fat, fat suppression MR imaging is well suited for these kind of tumors[10]. Unlike other vascular tumors, angiolipoma do not typically contain vascular flow on MRI. This may be due to presence of capillaries and venous channels which distinguish them from other lesion which predominantly contain arteriolar circulation .Lesions such as arteriovenous malformation usually show fast flow as flow void phenomenon[11].

Pathologically it is composed of mature fat cells with numerous small blood vessels. Thin fibrous capsule with fibrous septa usually divides the mass into lobules. Presence of fibrinous micro thrombi in the lumen is a diagnostic feature. The vascular component may be patchy and frequently accentuated in subcapsular area. The vessels are predominantly capillaries with some fibrin thrombi. Fibrosis may be associated with vascular component .The cellular variant includes angiomatous tissue which may be of spindle cells variety abundant in cellular areas and associated with mild pleomorphism. Numerous mast cells are sometimes seen throughout the tumor. Degenerative changes (i.e. hyalinization, myxoid change and fibrosis) may be present in longstanding cases. Mitotic figures are usually inconspicuous but if seen has no clinical significance[12]. Immunochemistry can be helpful especially CD34 and CD31 which stains endothelium of the blood vessels .The genetic studies have generally shown no abnormalities in the karyotype of these patients[13]. Angiomyxolipoma is a common differential diagnosis which is characterized by myxoid stroma. Unlike angiolipoma which has normal karyotype, angiomyxolipoma shares abnormal cytogenesis with lipoma, spindle cell lipoma and myxoma. Other common differential diagnosis is well differentiated angiosarcoma of the breast, Kaposi Sarcoma or primitive capillary hemangioma. The histogenesis is unknown but largely angiolipoma can be regarded as some sort of maldevelopment at embryonic time. The hypothesis of hamartomatous origin from primitive pluripotent mesenchymal cells is gaining wide acceptance. However others think it is neoplastic or hamartomatous diseases by origin. The more invasive variety may be a shift towards hemangioma end of this spectrum[14].

The main stay of definitive treatment is complete surgical extirpation. Non-infiltrating extradural angiolipoma are amenable to total resection but infiltrating angiolipomas are very difficult to remove entirely, especially if they are anterior or anterolateral in location[15]. It may then be necessary to undertake a staged removal, including an anterolateral approach with spinal stabilization. Since these lesions are quite slow growing and do not undergo malignant transformation, even with partial surgical resection patients get substantial symptomatic relief in terms of pain and neurological status. The role of external beam radiation is quite controversial. However there are three cases in literature out of 75, where postoperative radiation had been given following partial excision[16]. Angiolipomas are benign entities and even with partial surgical resection prognosis is quite good so any adjuvant radiation should be given with caution. Most of the authors contradict use of radiation in angiolipoma.

CONCLUSION

Spinal angiolipoma is rare but well defined clinical entity with distinguished histology and imaging features. The treatment for both infiltrating and non infiltrating angiolipomas is total surgical excision. Radiation therapy is not advocated even if there is some residual tumors.

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