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Unusual Presentation of Arterio-Venous Malformation Mimicking As a Soft Tissue Sarcoma

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Abstract:
Introduction: *Vascular malformations are congenital lesions due to abnormal embryonic development of vascular structures and they are subdivided into arteriovenous, capillary, venous, lymphatic and combined malformations.*
Case report : *A 16year old boy came to surgical OPD with complaints of swelling on the back on left side since 6 months. There was no history of trauma, fever. The swelling was insidious in onset, gradually progressive. It was initially of small size and increased to attain the present size. On examination, the swelling was noted in left thoraco-lumbar region approximately of size 10X8cm.The swelling was immobile. Skin was pinchable. No skin changes were noted. The swelling was firm in consistency.*
Discussion : *Intramuscular venous malformations are rare entities. They occurred most often in the head and neck and extremities but are relatively rare in the trunk and well localized to a single muscle or adjacent muscle groups.[4] Because venous malformations are lesions due to abnormal embryonic development, it is assumed that localized venous malformations result from insults of the specific neurovascular bundles during development, which is the origin of some localized vessels and muscles.*
Conclusion : *Arterio-venous malformations are rare entities which present in childhood. Delayed presentation of such conditions are very rare. In our case it was presented as a soft tissue sarcoma clinically which is rarest of rare.In conclusion, arterio-venous malformation presenting as a soft tissue sarcoma is very rare and few cases have been reported in the literature.*
Keywords: *arterio-venous malformation, intramuscular, soft tissue sarcoma*

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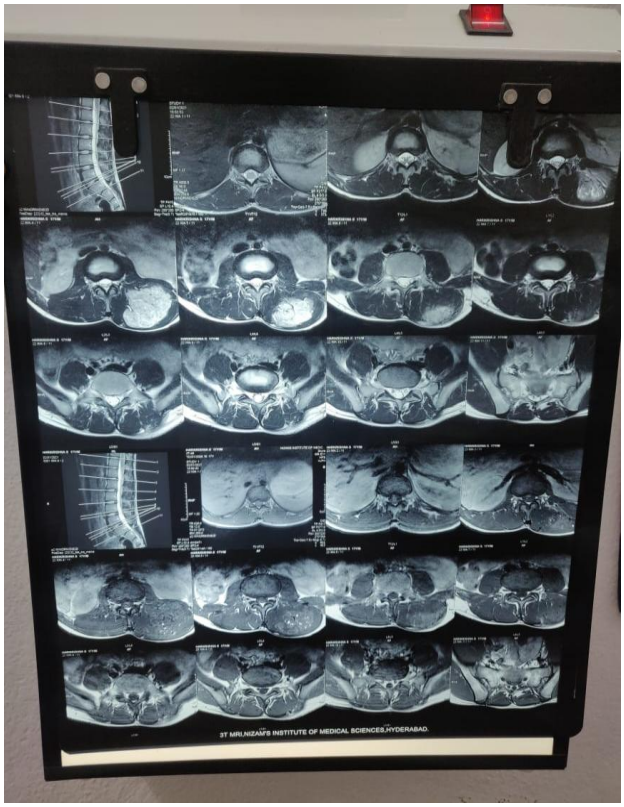
I. Introduction

Vascular malformations are congenital lesions due to abnormal embryonic development of vascular structures and they are subdivided into arteriovenous, capillary, venous, lymphatic and combined malformations.[1] Among them, venous malformations are the most common form and they are mostly in the skin and subcutaneous tissues.[2,3] In this report, with a review of literature, we describe a patient with extensive intramuscular arterio-venous malformation which involved paraspinal muscle group. Some previous cases reported a focal lesion of one or two adjacent muscles but extensive intramuscular arterio-venous malformation has rarely been reported.

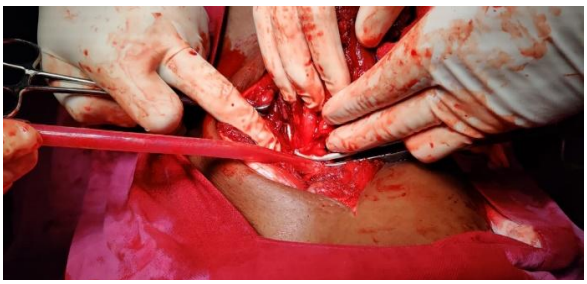
II. Case Report

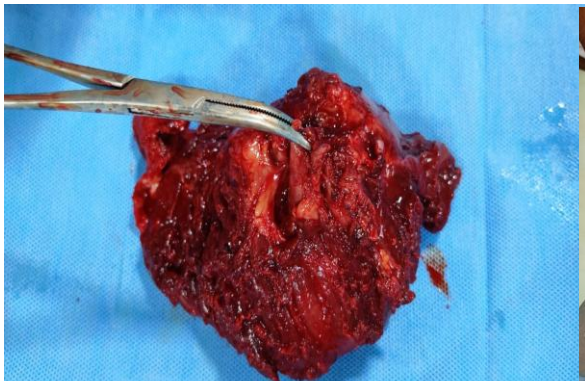
A 16year old boy came to surgical OPD with complaints of swelling on back on left side since 6 months. There was no history of trauma, fever. The swelling was insidious in onset, gradually progressive. It was initially of small size and increased to attain the present size. On examination, the swelling was noted in left thoraco-lumbar region approximately of size 10X8cm.The swelling was immobile. Skin was pinchable. No skin changes were noted. The swelling was firm in consistency. Initially it was thought to be of soft tissue origin. USG of the swelling reveals it to be a well defined encapsulated heterogenouspredominantly hyperechoic lobulated soft tissue mass measuring approx. 7.6X6X4.4cm noted in the intra/inter muscular plane of the back muscles at left thoraco-lumbar region with increased internal vascularity. The mass appears to be supplied by a single artery appears to be arising from intraabdominal region likely-aorta. CECT Abdomen and pelvis shows a mass in the left paraspinal muscles measuring approximately 5.4x5.4x10.2 cm as described above ? Sarcoma.

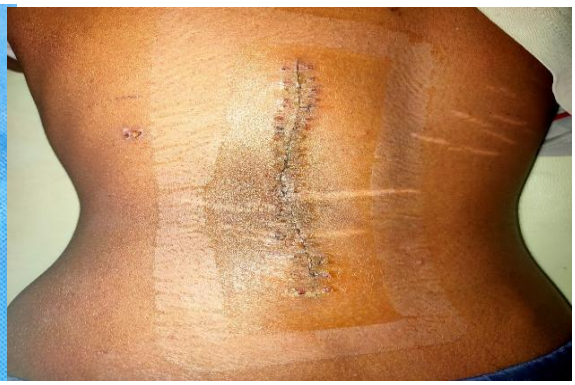
MRI of LS Spine shows lobulated altered signal intensity lesion in form of T1 hypointense with hyperintense foci within T2 & STIR hyperintense in left para spinal muscle displacing quadratus lumborum muscle anteriorly –Suggestive of tissue neoplasm.

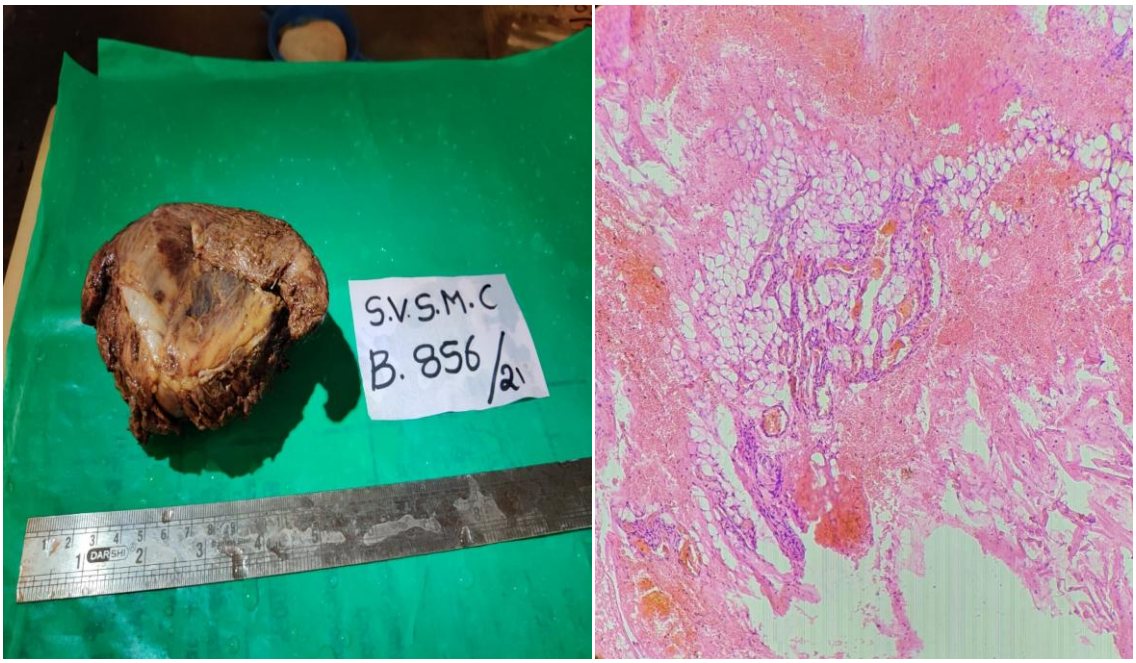


FNAC of the swelling reveals RBC & formed elements of blood only. Repeat FNAC was also not conclusive. Patient was taken up for surgery considering it to be a soft tissue sarcoma. Intraoperatively, before excising the tumour intoto , a syringe was introduced into the tumour and aspirated and found to be surprisingly blood. Later careful dissection was carried out to ligate the feeding vessels arising from left 2nd lumbar artery. The tumour was excised intoto and sent for histopathological examination. Cut section shows nodular grey white areas with focal grey brown areas .Irregular cut surface with pinpoint haemorrhages ,firm to hard with mucoid feel. Microscopic findings showed striated muscle fibres separated by vascular channels of varying calibre. Thick large channels,thin walled dilated channels and capillary proliferations are intermingled. Fatty infiltration into the stroma noted. No evidence of any sarcomatous lesion.









III. Discussion

Intramuscular venous malformations are rare entities. They occurred most often in the head and neck and extremities but are relatively rare in the trunk and well localized to a single muscle or adjacent muscle groups.[4] Because venous malformations are lesions due to abnormal embryonic development, it is assumed that localized venous malformations result from insults of the specific neurovascular bundles during development, which is the origin of some localized vessels and muscles. According to a study by Hein at al.[4] two-thirds of intramuscular venous malformation were also noted at birth and the remainder manifested in childhood and adolescence. However, it has the potential to be missed because they are frequently asymptomatic and their involved sites are invisible, especially during their early stages. In our patient, diagnosis of arterio-venous malformation was delayed until the age of 16 years, because the pain and pressure of the muscles were not triggered by movements in her daily life, and first appeared when he started yoga and stretching exercises. The superficial vascular malformations were thoroughly examined by ultrasound, with gray scale studies defining the extent and spectral and color doppler interrogation used to identify the flow characteristics.[5] Although the patient's arterio-venous malformation was identified by ultrasonography, an MRI is the most common and accurate tool of the early diagnosis of intramuscular arterio-venous malformation. But it is not the case in our scenario, MRI showed it to be a soft tissue neoplasm. Proper methods of treatment should be decided on after a full consideration of the degree of disabilities in daily living, injuries of adjacent tissues and cosmetic concerns. Recurrence, focal fibrosis or contracture following surgery is also more common with diffuse arterio-venous malformations. Our patient had minor symptoms and no disabilities in daily living for his lesion.

IV. Conclusion

Arterio-venous malformations are rare entities which present in childhood. Delayed presentation of such conditions are very rare. In our case it was presented as a soft tissue sarcoma clinically which is rarest of rare. In conclusion, arterio-venous malformation presenting as a soft tissue sarcoma is very rare and few cases have been reported in the literature.

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