Angiosarcoma of Manubrium Sterni

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Abstract
A rare case of angiosarcoma affecting the manubrium sterni is being reported. Angiosarcoma is a very rare tumour affecting the bones with incidence as less as 0.13% and its incidence involving the manubrium sterni is extremely rare.

Key Words
Angiosarcoma, Manubrium Sterni

Introduction
Angiosarcoma is a malignant tumor of endothelial histogenesis also known as angioendothelioma, it occurs most frequently in the skin, soft tissues, liver, kidney, spleen and reproductive organs (1). Angiosarcoma of bone at any site is quite rare. Mirra in a series of 3000 bone tumors found that the incidence of bony angiosarcomas was only 0.13% (1). Unni et. al. (2) at the Mayo Clinic found only one sternal angiosarcoma out of the 22 skeletal angiosarcomas.

Case Report
57 year male presented with swelling in the upper chest of 5 months duration gradually increasing in size. There was history of dull aching pain associated with this swelling however no history of fever, cough or hemoptysis was obtained. Patient looked well preserved. Examination revealed a hard swelling 6cmx3cm arising from the manubrium sterni which was not mobile and was noncompressible. Patient was investigated, skiagram of sternum revealed a bony lesion arising from the manubrium sterni, this finding was confirmed on CT scan, there was no infiltration of underlying structures. FNAC from the lesion was done which showed round cells in a haemorrhagic background.

Patient underwent surgery and on exploration there was a mass 6 × 6 cms. arising from manubrium sterni and the tumour was free from underlying structures and could be excised completely along with adjoining costal cartilages and the defect in sternum was replaced with prolene mesh and both ends of clavicle were stabilized with insertion of stainless steel plate (Fig 1). Postoperatively patient behaved well. Histopathology revealed tumour as low grade angiosarcoma (Fig 2).
Fig. 1. Post-operative x-ray chest PA view showing stabilisation of clavicles after excision of the tumour from manubrium sterni.

Fig. 2. Low power section showing features of a vasoformative tumour infiltrating and incarcerating skeletal muscle bundles and bony chips suggestive of low grade angiosarcoma.

Discussion

Angiosarcoma is a malignant tumour of endothelial cell histology, out of these, sternal angiosarcomas are very rarely reported. Mirra et al. has reported 3000 bone tumours with only 4 angiosarcomas comprising 0.13% (1). In other series at Mayo Clinic 1971 UNNI et al. described 22 skeletal angiosarcomas with only one in sternum (2). The lesions are characterised by solid sarcomatous areas, abortive vessel formation and vascular areas exhibiting anastomotic channels. Whereas in our case the section of the tumour showed a vasoformative tumour infiltrating and incarcerating skeletal muscle bundles and bony chips suggestive of low grade angiosarcoma. Another series of primary angiosarcomas of bone reviewed by Dorfman et al. (3), revealed 15 cases between 1930 to 1970, 9 lesions were located in the extremities 2 in the vertebrae and 2 in sternum. Angiosarcomas are usually primary, but there are reports of their occurrence as associated abnormalities. Abdelwahab et al. (4) described angiosarcomas occurring in bone infarct. Cutaneous angiosarcoma has been reported with metastasis to bone (5). Brady et al. (6), reported that less than 5% of all sarcomas, including angiosarcomas can be related to radiation exposure. Our case had no history of radiation exposure, there was no associated cutaneous angiosarcoma and in absence of bone infarct the lesion was considered as isolated primary angiosarcoma.

References