QUADRIAPAERESIS - A RARE CASE OF EPITHELOID HAEMANGIOENDOTHELIOMA.

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Abstract:
A 62 yr old male presented with complaints of neck pain associated with weakness of both upper and lower limbs for 2 months. Weakness began with upper limb progressing to involve the lower limbs. X-ray cervical spine showed C7 vertebral collapse with decreased joint spaces. MRI scan showed compression collapse of C7 vertebral body, thecal sac narrowing & cord changes at this level. C7 corpectomy and biopsy with anterior fusion with bone graft and instrumentation was performed revealing mass around C7 vertebra. Histology revealed fragments of cartilage, bone, & vascular tumor-composed of well formed vessels lined by cuboidal to hobnail endothelium. Focal areas showed lymphocytic infiltration and occasional benign giant cells suggestive of haemangioendothelioma. The purpose is to present a rare tumour occurring at an unusual site presenting as cervical myelopathy.

Keywords: Quadriapaesis, Haemangioendothelioma.

Introduction:
Haemangioendothelioma
The term represents a wide spectrum of vascular neoplasms.
Types –
1. Epitheloid hemangioendothelioma
2. Malignant haemangiosarcoma
Epitheloid hemangioendothelioma

Synonyms: low grade anaplastic angiosarcoma, cellular hemangioma, histiocytoid hemangioma angioendothelioma. They have well defined vascular channels. Tumor cells are plump & often cuboidal thus resembling epithelial cells. Occur in the calvarium, spine, femur, tibia and feet of adults during the second or third decade. Occurrence of this tumour in cervical spine is quite rare with best of our knowledge is 4th such case.

Case Report:
62 yr old male presented to out patient department on 26/12/2007 with complains of neck pain associated with weakness of both upper and lower limbs for 2 months. Pain was insidious in onset, gradually progressive, radiating to upper limbs.

Began with upper limb progressing to involve the lower limbs. Required support for mobility since then. For past 1 week has not been able to ambulate.


Discussion:
The term represents a wide spectrum of vascular neoplasms. Types are
X-ray cervical spine showed C7 vertebral collapse with decreased joint spaces. MRI scan showed compression collapse of C7 vertebral body, thecal sac narrowing & cord changes at this level.

Secondaries, tuberculosis were thought of clinically. USG abdomen was normal, routine haemogram was normal apart from ESR being 26 mm/hr. Sputum for AFB - negative, PSA - normal. C7 corpectomy and biopsy with anterior fusion with bone graft and instrumentation was performed revealing mass around C7 vertebra. Histology revealed fragments of cartilage, bone, & vascular tumor- composed of well formed vessels lined by cuboidal to hobnail endothelium. Focal areas showed lymphocytic infiltration and occasional benign giant cells suggestive of haemangioendothelioma. Postoperatively his vitals were stable, his pain had reduced motor deficits persisted with tingling numbness of fingers and toes. Short course of steroid was started on the second post op day. Patient was mobilized with walker on 3rd postoperative day. Was discharged with hard cervical collar.

X-ray shows expansile, osteolytic and poorly demarcated lesions with distinctive “soap-bubble” matrix with a sclerotic margin. There is no periosteal reaction. Aggressive lesions have ill-defined margins and loss of trabeculae. Treatment: Grade the lesion. Benign- curettage or wide resection. The role of radiation and chemotherapy is controversial. If Malignant - wide excision with additional radiotherapy/chemotherapy. Prognosis: Epithelioid hemangioendotheliomas may metastasize to bone and lung. Prognosis in these rare instances has been variable.
1) Epitheloid hemangioendothelioma

2) Malignant haemangiosarcoma Synonyms are low grade anaplastic angiosarcoma, cellular hemangioma, histiocytoid hemangioma, angioendothelioma.

Epitheloid hemangioendothelioma has well defined vascular channels. The tumor cells are plump & often cuboidal thus resembling epithelial cells. Occur in the calvarium, spine, femur, tibia and feet of adults during the second or third decade.

References: