Case Report-IV

Aggressive Sinonasal Hemangiopericytoma Presenting with Liver Metastasis : A Case Report

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ABSTRACT

Hemangiopericytoma is an uncommon soft tissue sarcoma. The tumour represents 2-3% of all soft tissue sarcomas in humans. This sarcoma arises from Zimmerman's pericytes. Sinonasal type hemangi-opericytoma is an uncommon tumour of the upper aerodigestive tract. The majority of sinonasal hemangiopericytoma behave in a benign manner with excellent long term prognosis. Here we report an aggressive case of sinonasal hemangiopericytoma presenting with liver metastasis. This 42 year old male presented with epistaxis, a recurrent mass obstructing the lumen of the nose and a lump in right hypochondrium. His diagnosis after biopsy was interpreted as hemangiopericytoma. Ultrasonography showed multiple metastasis in the liver which were also positive for metastatic sarcoma. This case highlights the fact that the usually benign sinonasal hemangiopericytoma can present with metastasis.

INTRODUCTION

Hemangiopericytoma is an uncommon soft tissue sarcoma and accounts for 2-3% of all soft tissue sarcomas in humans. This sarcoma arises from Zimmerman's pericytes, cells that are located around the blood vessels.[1] Sinonasal type hemangiopericytoma is a still uncommon tumour of the upper aerodigestive tract. The majority of sinonasal hemangiopericytoma behave in a benign manner with excellent long term prognosis.[1]

Here we report an aggressive case of sinonasal hemangiopericytoma presenting with liver metastasis.

CASE: A 42 year old male presented with one year history of epistaxis, blockage of nose and nasal twang. Six months ago he had undergone a surgical excision procedure at local hospital. However, treatment details were not available. Since one month before current admission was he had developed upper abdominal lump on the right side with upper abdominal pain and dyspepsia. On physical examination-the patient was anemic. There was a mass in the left nasal cavity which was completely obstructing the lumen of the nose, and bled on touch. The other general and systemic examination was unremarkable except for a firm , tender and nodular palpable liver. Invetigations: Blood-Hb. 5.7. Gm%, other haematological parameters were within normal limits. Bone marrow aspiration showed normocellular marrow with well preserved marrow components and there was no evidence of involvement of the bone marrow by any malignancy. Blood chemistry was within normal limits except for elevated

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liver enzymes- alkaline phosphatase as 649 IU/L, SGOT-142 IU/L SGPT-106 IU/L. Viral markers for hepatitis B and HIV were negative. Chest skiagram was normal.

CT Scan of paranasal sinuses showed a well defined lobulated sharply enhancing mass lesion in both nasal cavities extending into ethmoid air cells, sphenoid sinuses and nasopharynx (Fig 1). The lesion bulged into the left orbit and both

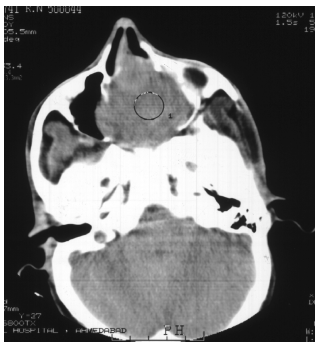

Figure 1 – CT Scan of paranasal sinuses showing the tumour

maxillary sinuses with retained secretions were seen in all paranasal sinuses. Ultrasonography of abdomen and pelvis showed liver enlargement with multiple hypoechoic areas in both lobes, largest measuring 121x98 mm in size, suggestive of multiple secondaries.

Histopathology report of biopsy from nasal mass in low power view (Fig 2) showed vascular spaces of the staghorn type, spindle shaped

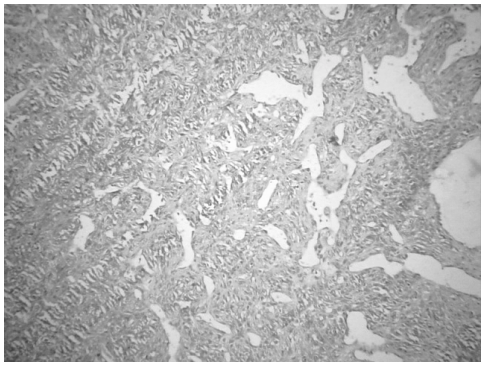

Figure 2 – Histopatholgical appearance of the nasal mass

plump tumour cells proliferating in sheets with high degree of cellularity. The high power view showed vesicular nuclei with nucleoli, evidence of giant cell formation, 6-8 mitotic figures per high power field. At places there was evidence of vascular invasion and thrombosis. Overall picture was interpreted as hemangio-pericytoma. US guided biopsy from hypoechoic liver lesions revealed plump to ovoid cell proliferation with hyperchromatic nuclei with adjacent normal liver tissue. The immunohisto-chemical profile showed the tumor cells reacting positively to vimentin, S-100, smooth muscle actin, muscle specific actin and factor XIIIa. Factor VIII was positive in vascular endothelial cells. CK, desmin and epithelial membrane antigen were absent. The conclusion was hemangiopericytoma.

We offered the patient palliative chemotherapy in the form of single agent adriamycin at a dose of 50 mg/m$^{2.}$

DISCUSSION

Hemangiopericytoma is primarily a tumour of adults and is rare in infants and children. There is a long natural history; peak incidence of tumour is around 6th decade of life. It occurs equally in both sexes.[1] Theoretically, hemangiopericytomas can occur anywhere where there are blood vessels. The tumour is most common in lower extremity especially thigh, pelvic fossa and retroperitoneum. Most are deep seated and the majority are found in muscle tissue.[1] Hemangiopericytomas are reported to occur at many other sites in the body including intraosseous,[1] nasal cavity and sinuses (sinonasal)[2] meninges.[3] It is a lesion which is benign but definitely has a malignant counterpart. Retroperitoneal and meningeal tumours had a higher local recurrence rates when compared to extremity tumours which fare significantly better.[4] Histological features of aggressive biological behaviour are presence of necrosis, mitoses, vascular invasion and pleomorphism. High grade lesions are the ones which usually metastasize, the more common site of metastasis being the lungs, metastasis also

occurs to the bones with lymph nodes and liver being the less common sites of metastasis.[5] As with other sarcomas these can present with hypoglycemia due to the secretion of IGF-I and IGF-II.[6] Lesional cells are reactive for vimentin, actin, factor XIII a, laminin and CD 34. In contrast to interspersed endothelial cells the lesional cells will not stain for factor VIII. Multiple chromosomal abnormalities have been observed including t(12;19) and t(13;22).[1] Treatment is primarily surgical with adjuvant radiotherapy for tumours in location eg retroperitoneal and meningeal tumors which have a higher recurrence rates.[4] Chemotherapy can help in achieving partial or complete responses in metastatic hemangiopericytomas. The drugs used are actinomycin-D, adriamycin, cyclophosphamide, methotrexate and vincristine.[5]

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