

CASE REPORT

INTRACRANIAL EXTRA-SKELETAL MESENCHYMAL CHONDROSARCOMA

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Intracranial Mesenchymal Chondrosarcoma is a very rare and uncommon entity that affects young adults. We came across one such patient who presented with severe headache and intermittent nausea and vomiting. The clinical, radiological preoperative diagnosis was a meningioma, on histological examination it turned out to be mesenchymal chondrosarcoma of tentorial region in posterior fossa, uncommon site for this entity.

Keywords: Mesenchymal chondrosarcoma, Tentorium, Meningioma

INTRODUCTION

Mesenchymal chondrosarcoma was firstly described by Liechtenstein and Bernstein¹ in 1959 as a special variant of chondrosarcoma. This tumour has got a characteristic histological diamorphic picture, sheets or cords of undifferentiated small round stromal or mesenchymal cells interspersed with islands of well-differentiated cartilage.² This is a rare neoplasm, originally described in bone,³ has occasionally been reported to arise intracranially attached to the dura, spinal meningies and cauda equina in childhood and young adults.⁴

We diagnosed a case of extraskeletal mesenchymal chondrosarcoma of right petrosal and tentorial region in posterior fossa, a rare site not reported before, on histopathology, at Gulab Devi Hospital, Lahore. The case is reported and the available literature is reviewed.

CASE REPORT

A young lady of 26 years was admitted with history of severe headache, intermittent nausea and vomiting for the last one and half year. Sometimes it causes discomfort particularly during sleep. There was no history of trauma, fever or weight loss. Her general physical examination revealed mild pallor. Neurological examination revealed right optic nerve swelling. Laboratory investigation revealed an ESR of 30 mm at the end of 1st hour and haemoglobin of 9.5 g/dl. X-rays skull revealed a soft tissue mass in the posterior fossa.

Contrast enhanced CT showed lobulation and solid tumoural enhancement with central low – attenuation areas. MRI showed a large, instantly enhancing mass measuring 5.5x4x3cms which appears to originate from the right side petrosal in posterior fossa with significant mass effect. After the clinical and radiological diagnosis of intracranial tumour, surgical intervention was planned.

On gross examination, the material revealed multiple, firm, dark brown pieces of tissue, weighing 8gms and varying from 2cm to 0.4cm in size,

(7x6x1cm in aggregate). Six representative tissue pieces were processed in automatic tissue processor (Shandon, England), embedded in paraffin and 3 to 5 micron thick sections were cut. The sections were stained with hematoxylin and eosin.

Light microscopic examination showed highly cellular and undifferentiated mesenchymal cells and island of well – differentiated cartilage (Fig-1). There are small whorls of some what fusiform cells and densely cellular small cell aggregate with areas of chondromatous differentiation (Fig-2). The undifferentiated mesenchymal cells were uniform, small with rounded hyperchromatic nuclei and hardly any cytoplasm. These cells are arranged in better pattern around thin walled blood vessels. Areas of chondrosarcoma are also present with tumour necrosis.

Fig-1: Mesenchymal Chondrosarcoma with cartilage and small blue cells X20

Fig-2: Mesenchymal Chondrosarcoma with well differentiated cartilage intermingled with small deeply stained malignant cells X40.

DISCUSSION

The salient feature of the case under review was the occurrence of a mesenchymal chondrosarcoma in the posterior fossa of a girl of 26 years. Mesenchymal chondrosarcoma is a very rare tumour, approximately 20.2% of all the primary bone tumours, but very rare in extra-skeletal tissue.⁵ The tumour is much more common in the 2nd decade⁴ or in the 3rd decade (3%) as reported by Mirra.³ Size of the tumour varies from 4 to 30cm (average 12.5cm).⁶ In the present case the size of the tumour was 5.5x4x3cm.

Mesenchymal chondrosarcoma has got a very distinctive histo-morphological picture. On the basis of the morphology these tumours are generally divided into two varieties – haemangiopericytoma variant as was seen mostly in this case and the less differentiated small cell type. Mesenchymal chondrosarcoma is to be differentiated from other soft tissue sarcomas namely malignant haemangiopericytoma, chondrosarcoma and synovial sarcoma.^{7,8} Focal distribution of cartilaginous tissue along with undifferentiated mesenchymal cells favours the diagnosis of mesenchymal chondrosarcoma the cartilaginous component is more pleomorphic and cellular with bizarre nuclei and mitoses.⁹ Dual epithelial and spindle cell elements are characteristic of synovial sarcoma.⁸

Genesis of the tumour is obscure. However, the transition areas between the spindle shaped mesenchymal cells and cartilage islands clearly indicate that the tumour takes its origin from the precartilaginous mesenchyme.¹⁰

Surgical resection and radiation therapy to the tumour bed is recommended where the tumour is

resectable, chemo therapy is the treatment of choice in recurrent cases. A long term prognosis is poor.

REFERENCES

1. Lichten Stein L, Bernstein D. Unusual benign and malignant chondroid tumours of bone. A surgery of some mesenchymal cartilage tumours and malignant chondroblastic tumours, including a few multicentric ones, as well as many a typical benign chondroblastomas and chondromyxoid fibromas. *Cancer* 1959; 12: 1142-1157.
2. Scheuthaner BW, Rubinstein LJ. Meningeal mesenchymal chondrosarcoma: report of 8 cases with review of the literature. *Cancer* 1978; 42: 2744-52.
3. Mirra JM. Bone In: "Surgical Pathology: WF Coulson, editor. Philadelphia: JB Lippincott Co; 1988, pp 1356-1471.
4. Rushing EJ, Mena H, Smirniotopoulos JG. Mesenchymal Chondrosarcoma of the cauda equine. *Clin Neuropathol* 1995; 14: 150-3.
5. Tyagis, Maheshwari V, Tyagi N. Mesenchymal chondrosarcoma a case report. *The Postgrad med* 1992; 38: 34-5.
6. Naumann S, Krallman PA, Unni KK, Fidler ME, Neff JR, Bridge JA. Translocation der (13; 21)(q10: q10) in Skeletal and Extra skeletal Mesenchymal chondrosarcoma. *Mod Pathol* 2002; 15:572-6.8.
7. Kleist B, Poetsch M, Lang C, Bankau A, Lorenz G, Suess- Fridrich K et al. Clear cell chondrosarcoma of the Larynx, A case report of a rare histologic variant in an uncommon localization. *Am J Surg Pathol* 2002; 26: 386-392.
8. Tsunneyoshi M, Daimaru Y, Enjoji M. Malignant hemangiopericytoma and other sarcomas with hemangiopericytoma like pattern. *Pathol Res Pract* 1984; 178: 446-53.
9. Aigner T, Oliveira AM, Nasiemento AG. Extra skeletal myxoid chondrosarcoma do not show a chondrocytic pheno type. *Mod Pathol* 2004; 17: 214-21.
10. Mc Grory JE, Rock MG, Naseimento AG, Oliverira AM. Extra skeletal myxoid chondrosarcoma. *Clin Ortho P* 2001; 382:185-90

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