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Intracranial extradural low-grade fibromyxoid sarcoma presenting as proptosis in a Human immunodeficiency virus (HIV) positive patient

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ABSTRACT

Background: Low-grade fibromyxoid sarcoma(LGFMS) are rare mesenchymal tumors that have a benign histology with a potentially aggressive clinical behavior and a high rate of recurrence and metastasis.

Case Description: We report a case of a 26-year-old female known HIV positive on Antiretroviral therapy (ART) who presented with protrusion of left eyeball since six months, initially gradual and painless, followed by a rapid course of progressive proptosis and chemosis associated with severe headache and repeated episodes of vomiting for twenty days. The imaging was suggestive of a large intracranial extradural locculated Cerebrospinal fluid (CSF) attenuating mass lesion with left intra orbital extension. After all baseline investigations she underwent left frontotemporal craniotomy and tumour excision. Histological examination of the lesion was suggestive of a Low grade fibromyxoid sarcoma (LGFMS).

Discussion: LGFMS are slow-growing benign soft-tissue tumors which are usually asymptomatic and have a high rate of local recurrence and late metastasis. Central Nervous System (CNS) involvement in HIV positive patients comprises of infections, tumors (Primary CNS lymphomas, Metastasis, Kaposi Sarcoma) and other conditions such as Progressive Multifocal Leukoencephalopathy, Dementia and Neuropathies. However such kind of tumor in HIV positive patients hasn't been reported in literature. Also in our case, apart from the histological rarity, the location of this tumor was also highly unusual.

Conclusion: There are rare reports of such intracranial tumor in adults, however none of them presented with proptosis due to an extradural infiltrating LGFMS . To the best of the author's knowledge, this is the first reported case of this rare disease in a HIV positive patient with an even rarer location and presentation.

Keywords: fibromyxoid sarcoma; intracranial; HIV; proptosis ; extradural

INTRODUCTION

Low-grade fibromyxoid sarcoma (LGFMS) is a mesenchymal, soft tissue-tumor that is characterized by a benign histological appearance of contrasting fibrous and myxoid areas, a swirling, whorled growth pattern, and bland, deceptively benign–appearing fibroblastic spindle cells but an aggressive clinical behavior.[1,2] The most common anatomical locations are the lower extremities, thorax, inguinal area, and upper limbs as a painless enlarging mass.[3] There are very few prior cases reported in literature describing intracranial involvement in adults and one case in a child till 2018 [4-6] However ,to our knowledge, intracranial extradural involvement of LGFMS in a HIV positive patient with a presentation of proptosis has not been previously described in literature. We report our experience and management of such a case on retroviral therapy with a rapidly expanding intracranial extradural lesion who presented to us with rapidly progressing proptosis.

Case Report

History: A 26 year old known HIV positive female on retroviral treatment presented with history of progressive protrusion of left eyeball for six months (Figure 1A). It was insidious in onset, gradually progressive and painless. It was associated with

moderate throbbing headache and intermittent vomiting. She was then referred to the department of neurosurgery and was advised admission but the patient refused and went to another centre for a second opinion.

She again presented to us after twenty days with complaint of increasing severe protrusion of the left eyeball with reddening, edema, pain (Figure 1B), increase in headache and vomiting. However there was no history of any double vision and no other manifestation suggestive of Acquired Immunodeficiency syndrome (AIDS).

Examination:

On examination there was asymmetrical proptosis of the left eye of 3cm with outward and downward displacement of eyeball. The inferior orbital wall margin was not palpable with superior sulcus fullness was present. Prominent blood vessels and redness on skin with lid crease was absent along with the presence of lagophthalmos. She had mild congestion along with mild exposure keratopathy on the first visit (Figure 1) however there was presence of severe congestion with severe exposure keratopathy with dry eye and hazy cornea (Figure 2). On auscultation bruit was absent. Fundus examination revealed papilledema with dull foveal reflex.

Imaging:

The Non-contrast computed tomography (NCCT) head and orbit was suggestive of a large 5*7*6.7 cm locculated hypodense extraaxial mass lesion with peripheral rim of calcification with intra orbital extension into ipsilateral intraconal space through inner table of left frontal bone and superior orbital fissure without any evidence of globe or optic nerve involvement. There was a significant proptosis seen on the left side along with marked thinning of the left anterior cranial fossa floor and frontal bone without any destruction or invasion. Marked expansile scalloping of left orbital roof was seen causing downward compression and anterior displacement of the extraocular muscle cone and left orbit resulting in severe proptosis. (Figure 2)

Magnetic resonance imaging (MRI) Brain was then done which revealed a large extradural frontotemporal cystic solid tumor which was hypointense on T1 and hyperintense on T2 weighted images along with partial suppression on FLAIR images. There was no diffusion restriction seen. A caudal solid nodule was seen along the medial aspect of lesion at roof of left orbit showing profound gradient susceptibility. Post contrast images revealed an amorphous nodular solid enhancement of this portion along with minimally enhancing fine reticulations within the cystic lesion (Figure 3). MR angiography and B-Scan of both eyes were also done which were normal.

Management:

The patient underwent routine hematological investigations as per pre anaesthetic workup which were found to be in a normal range. Her CD4 counts were also done which were found to be 338 cells/microliter. After an informed consent and with universal precautions, the patient underwent left frontotemporal craniotomy with tumor decompression along with a temporary left eye tarsorrhaphy. Intraoperatively the tumour was a found to be large, well-defined, greyish brown, glistening extradural, mucoid filled with a gelatinous consistency along with some bony pieces (Figure 4).

Pathological findings:

Histopathology of the lesion was suggestive of fibromyxoid tissue with nerve fibers along with focal areas of bony trabeculae. The lesion appeared more fibrous than myxoid and had low cellularity composed of spindle shaped cells with small hyperchromatic oval nuclei with clumped chromatin. The cells showed mild nuclear pleomorphism with little mitotic activity (Figure 5). Also the specimen was positive for vimentin and negative for cytokeratin, EMA and S100 protein.

Post-operative course:

Post operatively, the patient remained stable and was discharged after seven post-operative days without any new deficits. She is under regular follow up and her ART was resumed on discharge and has not shown signs of any clinical deterioration and is undergoing the advised postsurgical adjuvant radiation therapy. A follow up MRI Brain is planned after three months.

Discussion

LGFMS was first described by Evans [1] in 1987 as slow-growing soft-tissue tumors which are usually asymptomatic and have a benign histology, but are

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quite deceptive due to a high rate of local recurrence and late metastasis. According to literature this tumour occurs in young to middle-aged adults, with the most common site being the lower extremities, followed by the upper extremities, chest wall, and abdominal cavity. As per our knowledge only six cases have been reported of intracranial involvement, five in adults and one cases in children [4-9].

Central Nervous System (CNS) involvement in HIV positive patients is not uncommon with infections (Cryptococcosis, Toxoplasmosis, Tuberculosis etc), tumors (Primary CNS lymphomas, Metastasis, Kaposi Sarcoma) and other conditions such as Progressive Multifocal Leukoencephalopathy, Dementia and Neuropathies amongst the familiar ones[10].

However, the report of LGFMS in HIV positive patients has not been made in literature and this is the first case of this rare disease in such a patient with an even rarer extradural location and presentation as progressive proptosis.

Due to the histological mimics, the pathological diagnosis of LGFMS is difficult and challenging, Myxomatous meningioma, chordoid meningioma are the most common differential diagnosis but they are positive for cytokeratin and EMA which wasn't in our case [4]. The other differentials include Myxoid chondrosarcoma, liposarcoma, and myxoma. Chondrosarcomas are positive for vimentin and S-100 , liposarcomas are positive for smooth muscle actin, along with vimentin and S-100 and myxomas are locally noninvasive [4,11-15]. Thus the presence of contrast fibrous and myxoid areas, swirling growth pattern, low to moderate cellularity, and slight nuclear polymorphism, as well as positive staining for vimentin without positive staining for cytokeratin, S100 protein, EMA, smooth muscle actin, and CD34 all supported the diagnosis of lowgrade fibromyxoid sarcoma [13,16,17].

Molecular analysis reveals the presence of ring chromosomes which is due to two highly specific translocations which are highly sensitive and specific for LGFMS. One of them is the fusion of the platelet-derived growth factor *(PDGF)* gene to a collagen A1 gene via the translocations from chromosomes 17 and 22. The other is the fusion gene of *FUS-CREB3L2* or *FUSCREB3L1* resulting from t(7,16) (q32–34;p11) or t(11,16) (p11;p11)[18].

The treatment of choice is surgery, and the prognosis is dependent on the extent of resection [1,2,18,19]. Surgery also remains the gold standard for local recurrence and for metastatic lesions. In the case reported by Saito et al the patient had recurrence six times and underwent treatment by surgical removal four times [4]. Also it is strongly recommended to follow the Universal Precautions strictly to prevent the feared spread of HIV infection to the operating surgeons and other attending medical personnel while dealing with such cases [20].

Authors have also reported treatment with chemotherapy or radiation, but it is not definitive that the adjuvant therapy tends to change the outcome of the disease or the recurrence rate or metastasis [4,11-13]. Nevertheless, radiation therapy including stereotactic radiosurgery contributed to tumor control in the case reported by Saito et al [4].

Proper counseling regarding the probability of recurrence must be explained and thus, long-term follow-up is a must. Our patient is under regular follow up and hasn't deteriorated clinically and is planned for a post-operative MRI at three months. The resumption of ART as early as possible is also essential to maintain the immune status and prolong life expectancy.

Conclusions

This present case of LGFMS with an intracranial extradural involvement in an individual of AIDS presenting with rapidly progressive proptosis is an extremely rare entity. It is a benign but aggresive tumour, treatment of choice of which is total resection. It has a high recurrence rate and thus a good prior counseling as was done in our case and long term follow up is essential for its management.

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Figure Legend:

Figure 1 (A): Clinical photograph of the patient on the first visit showing significant Left proptosis
(B): Clinical photograph of the patient on the second visit after 20 days showing worsening of the proptosis with severe congestion, chemosis and exposure keratopathy.

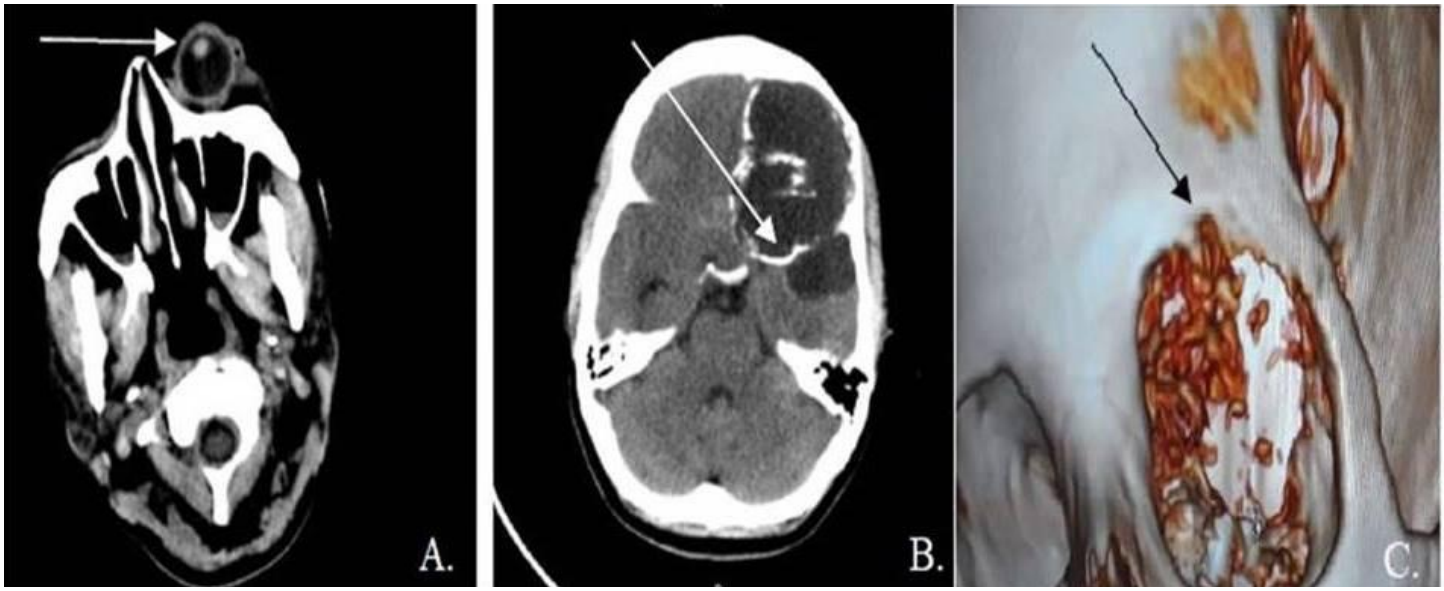

Figure 2: Non Contrast CT scans Head showing (A.) Proptosis (B.) A hypodense extraaxial mass lesion with peripheral rim of calcification (C) Marked expansile scalloping of left orbital roof

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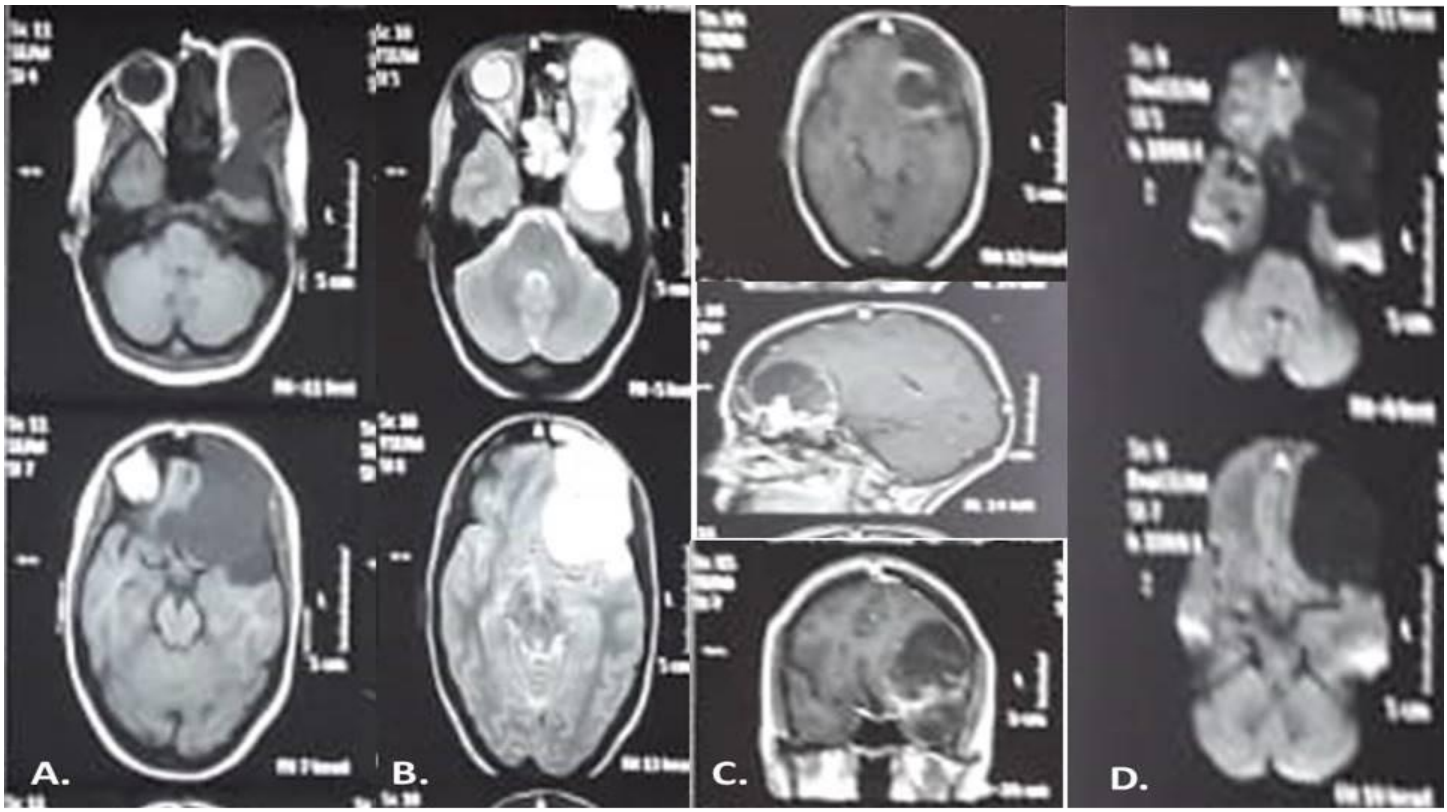

Figure 3: MRI brain with (A.) T1 weighted image showing hypointense lesion (B.) T2 weighted images showing hyperintense lesion (C.) Contrast enhanced images showing nodular solid enhancement of the medial peripheral part of the lesion along with minimally enhancing fine reticulations within the cystic lesion. (D.) Diffusion weighted images showing no restriction

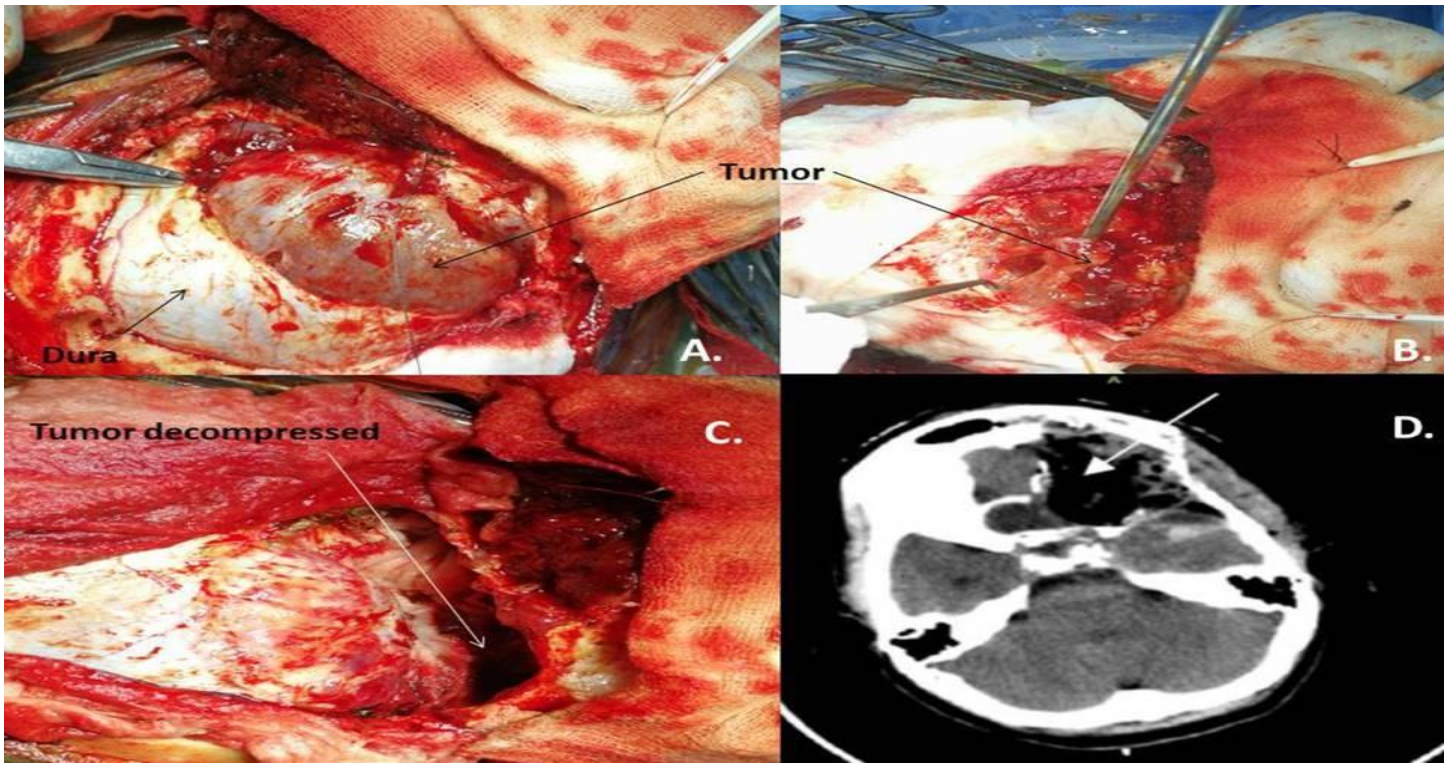

Figure 4: (A, B) Intraoperative extradural greyish brown lesion with a mucoid consistency
(C.) Post tumour excision picture showing adequate decompression (D) Post operative CT scan

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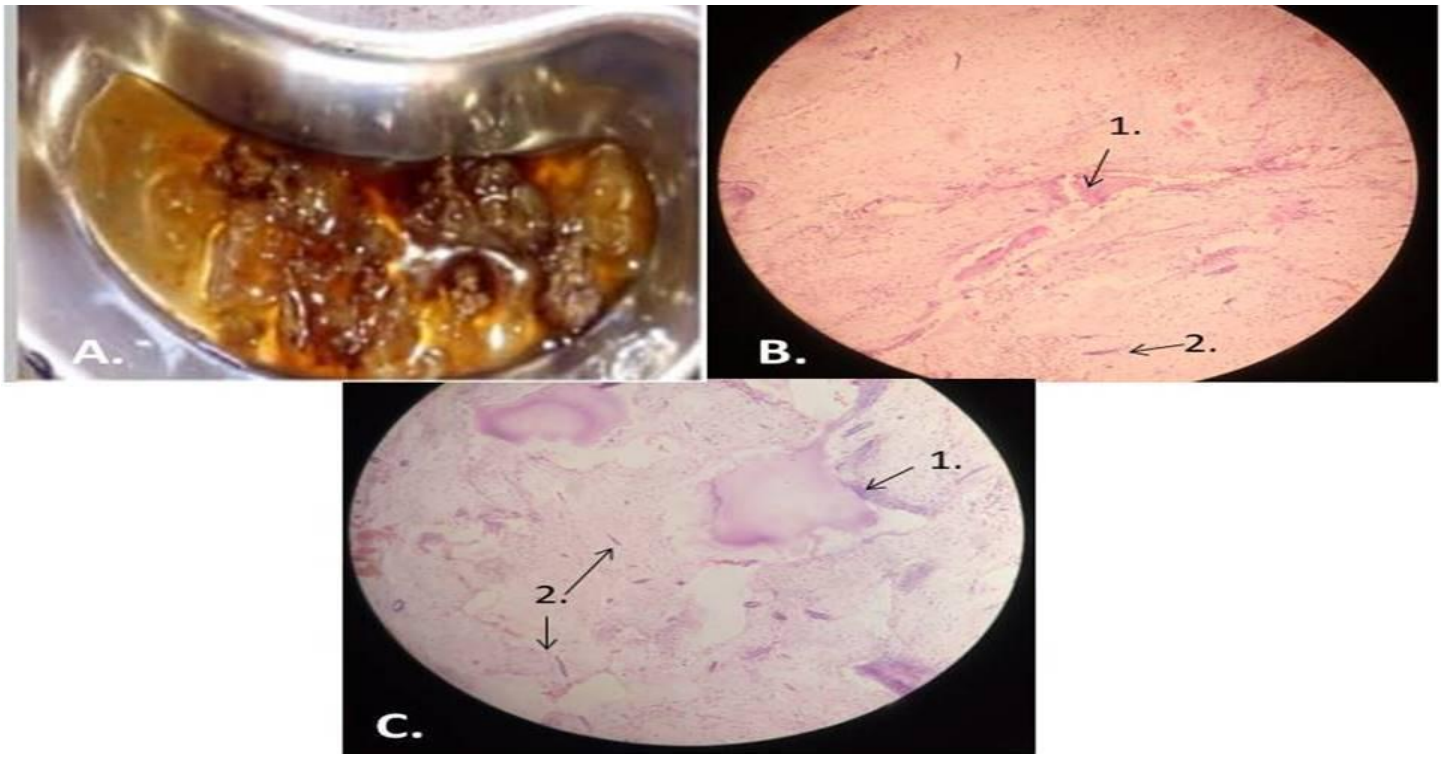

Figure 5: Histopathology slides (A.) Gross specimen of a greyish brown gelatinous tumour (B) Microscopic specimen (Hematoxylin and Eosin stain – 4x) showing 1. Myxoid degeneration and 2. Spindle cells
(C) Microscopic specimen (Hematoxylin and Eosin stain – 10 x) showing 1. Bony trabeculae and 2. Spindle cells

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