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CASE REPORT

Primary Intracranial Myxoid Chondrosarcoma: A case report

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ABSTRACT

Objective: Intracranial extraskeletal myxoid chondrosarcoma is a very rare tumor; up to date, only ten cases have been reported.

Case report: We present a case report of a 65-year-old female admitted to Zagazig university hospitals by right side hemiplegia of gradual onset and progressive course over three weeks. Magnetic resonance imaging (MRI) revealed left intra-axial parieto-occipital space occupying lesion, multiloculated, hypodense with central necrosis, heterogeneous enhancement, minimal brain edema, and causing a mass effect. The patient underwent subtotal resection using sonar guided craniotomy. The histopathological and immunohistochemical evaluation confirmed a diagnosis of extraskeletal myxoid chondrosarcoma. One month after initial discharge, the patient underwent radiotherapy. About 6 months after surgery, the patient was deteriorated and died.

Conclusion: Intracranial extraskeletal myxoid chondrosarcoma is a rare malignant cartilaginous tumor. Pathological diagnosis is the gold standard and radical excision is the standard treatment.

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KEYWORDS

Intracranial tumor; Chondrosarcoma; Central nervous system

Introduction

Primary intracranial extraskeletal chondrosarcoma is a rare neoplasm of the central nervous system representing <0.16% of the primary intracranial tumors [1]. The tumor usually arises from the base of the skull, choroid plexus, dura matter and brain parenchyma [2].

These tumors are potentially aggressive, invading the brain parenchyma and elevating intracranial pressure. Various symptoms were noted in these patients, although diplopia and headache are the commonest clinical symptoms [3].

Three histological variants of chondrosarcoma have been presented: Myxoid, mesenchymal and classic chondrosarcoma [4]. Few cases of primary intracranial extraskeletal myxoid chondrosarcoma have been reported up till now [5].

Case report

A 65 years old female was admitted to Zagazig University Hospitals at Neurosurgery department with right side hemiplegia of gradual onset and progressive course over three weeks associated with a blurring of vision, vomiting, and deterioration of consciousness four days before admission. Glasgow coma scale on admission was 12/15. Laboratory tests including complete blood count, liver, and kidney function tests were normal. The patient had no history of trauma or family history of hereditary illness or similar conditions.

On imaging, Axial MRI with contrast showing left intra-axial parietooccipital space occupying lesion, oblong in shape measuring 60×39×40 mm, multiloculated, hypodense with central necrosis, heterogeneous enhancement, minimal brain oedema and causing a mass effect (Fig. 1). Based on the preoperative imaging, left parietooccipital metastatic carcinoma or intracranial abscess was suspected. The patient underwent subtotal resection using sonar guided craniotomy due to its large size, and deep infiltration of the brain substance.

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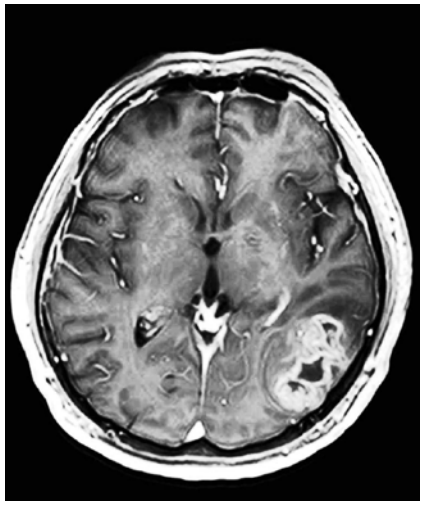

Figure 1. MRI brain with contrast axial cut. Showing left pareitooccipital intraxial space occupying lesion, oblong in shape with central necrosis, heterogeneous enhancement and minimal brain oedema.

Intra-operative gross examination of the tumor mass revealed intra-axial grayish white mass, gelatinous to firm in consistency with low vascularity invading the surrounding brain parenchyma.

On histopathology, sections show a characteristic multinodular pattern which was clearly evident at low magnification. The individual tumor nodules consist of round or slightly elongated cells of uniform shape and size separated by focal areas of fibromyxoid changes. The individual cells have small hyperchromatic nuclei and a thin rim of deeply eosinophilic cytoplasm reminiscent of chondroblasts. Characteristically, the individual cells are arranged in short anastomosing cords (Fig. 2).

On immunohistochemical examination, the tumor cells were negative for glial fibrillary acidic protein (GFAP, 1:200, Biogenex, San Ramon, CA, USA), EMA (epithelial membrane antigen (1:25, Dako, Glostrup, Denmark), pan-cytokeratin (AE1/AE3, 1:40, ab27988, Abcam, Cambridge, UK) and positive for MAP2 (microtubule-associated protein 2, 1:200, clone AP18, Neomarkers, Fremont, CA, USA), Vimentin (anti-vimentin antibody; ab52942; 1:200; Abcam, Cambridge, UK) and S100 (anti-S100 antibody; ab66041; 1:200; Abcam, Cambridge, UK) (Fig. 3). Thus, a pathological diagnosis of primary intracranial extraskeletal myxoid chondrosarcoma was confirmed. Postoperative cranial MRI identified subtotal resection of the tumor. The patient was discharged on the 8th postoperative day with relieving of some neurological deficits.

One month later, the patient was given 3D conformal radiation therapy 60 Gy/30 fractions/6 weeks 5 fractions per week on two phases; phase 1:44 Gy for tumor bed plus 2 cm margin, and phase 2:16 Gy to the residual tumor + 1.5 cm margin. This patient died 6 months postoperatively. Informed consent was obtained for the publication of this study.

Discussion

Intracranial myxoid chondrosarcoma is a rare neoplasm, which may arise from the dura, choroid plexus, or the pineal region [6,7]. Up to date, only ten cases of primary intracranial extraosseous myxoid chondrosarcoma have been reported [5]. Most of this rare neoplasm shows a dural involvement; except in very rare cases where it arises within the brain parenchyma without attachment to the meninges or the cranium as found in our case report.

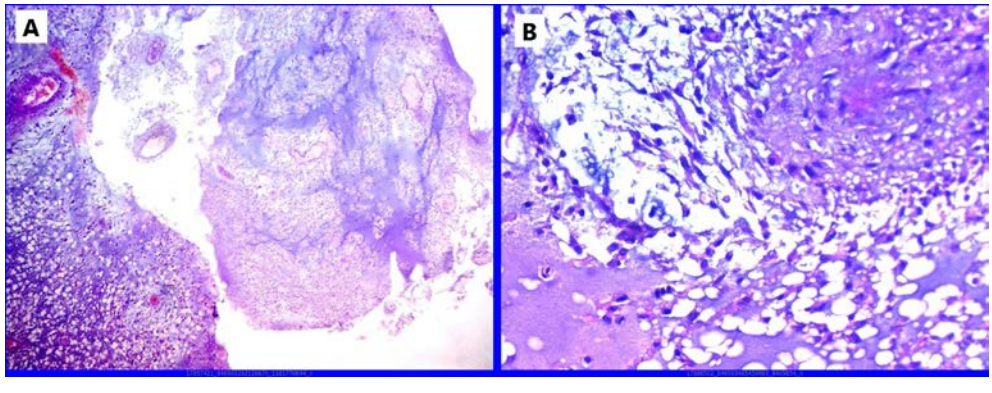

Figure 2. A: Histologic evaluation of this mass revealed a multinodular pattern that consists of an undifferentiated round or spindle-shaped cells with mature cartilaginous tissue and abundant myxoid stroma (H&E, ×100). B: The tumor cells interconnected to form cords and have small hyperchromatic nuclei and a narrow rim of deeply eosinophilic cytoplasm reminiscent of chondroblasts (H&E, ×400).

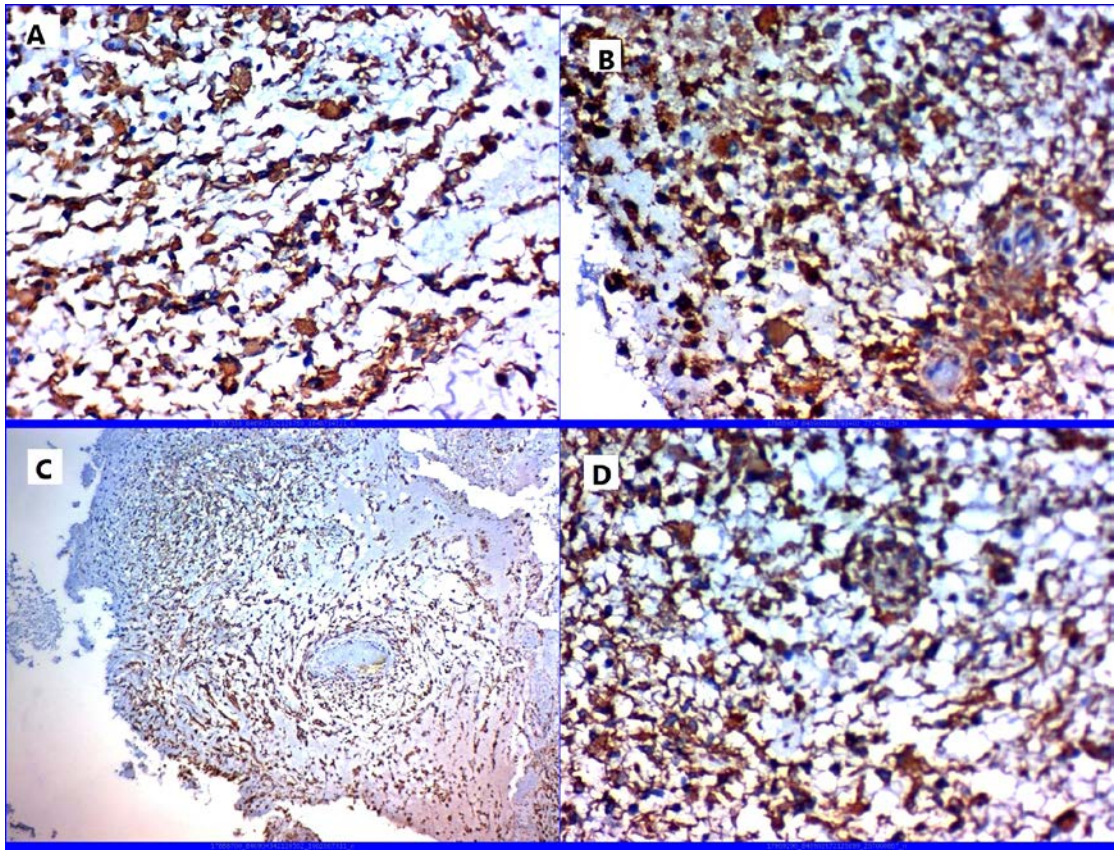

Figure 3. Immunohistochemical evaluation revealed tumor cells that are positive for S100 (IHC ×400) (A), microtubule-associated protein 2 (MAP2) (IHC, ×400) (B), Vimentin (IHC ×100) (C), Vimentin (IHC ×400) (D).

In this report, a case of myxoid type intracranial chondrosarcoma that occurred in the left parietooccipital region of the skull was presented, which has rarely been reported in the literature. With the help of immunohistochemistry; vimentin, cytokeratin, and S100, the chondrosarcoma can be differentiated from chordoma. Chordomas lack vimentin and chondrosarcomas don't express cytokeratin. S-100 protein expression is present in both. S100 is used as the major tool to determine neural crest differentiation and is frequently deployed to a spindle cell neoplasm of uncertain differentiation. S100B is more abundantly expressed in glial cells, melanocytes, adipocytes, and chondrocytes. S100 protein antibodies are often polyclonal but are typically strongly reactive against S100B [8].

The best treatment for this rare neoplasm is the radical excision. Adjuvant therapies, including radiotherapy, and proton beam treatment, that have been found to improve the patient outcomes [9]. Unfortunately; it was not associated with a good prognosis even after the treatment.

In conclusion, intracranial extraskeletal myxoid chondrosarcoma is a rare malignant tumor. Due to its rarity and similar imaging findings with meningioma and metastatic carcinoma, a differential diagnosis is often challenging and pathological diagnosis is the gold standard.

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