

A Rare Variant of Meningioma: Case Report and Review of Literature

Abstract

Meningioma is a morphologically heterogeneous tumour arising from meningeothelial cells that has been classified by the World Health Organization into 15 different histological types and graded into three types groups (Grade I, II, and III) based on the biological behavior. Metaplastic meningioma is a rare subtype of meningioma characterized by focal or widespread mesenchymal differentiation in the form of bone, cartilage, fat, or xanthomatous tissue elements. Xanthomatous meningioma is a subclass of metaplastic meningioma which is exceedingly rare. Only a few cases have been reported in the literature. Here, we report the case of a 44-year-old man, who presented with left sided weakness and was diagnosed as a case of xanthomatous meningioma.

Keywords: Meningioma, metaplastic, xanthomatous

Introduction

Meningioma is a group of slow growing neoplasms derived from the meningeothelial cells of the arachnoid layer. Metaplastic meningioma is one of the variants of benign meningioma with focal or widespread mesenchymal components. Xanthomatous meningioma is a rare subtype of metaplastic meningioma. Imaging features may point towards the diagnosis of xanthomatous meningioma, however definitive diagnosis requires histopathological examination. The prognosis in patients with this type of meningioma appears to be excellent. Here we report the case of a 44 year old man, who presented with left hemiparesis secondary to an extra axial space occupying lesion, which was diagnosed as a case of xanthomatous meningioma.

Case Report

A 44-year-old male presented with progressive weakness of left upper and lower limbs for 1 month. There was no history of headache, vomiting, or seizures. He did not have any other comorbid illnesses, addictions, allergies, or significant family history. On examination, his vital signs were stable. He was fully conscious and his cranial nerve examination was normal. He had Grade 4/5 power in the

left upper limb. The power in the left hip and knee was 3/5 while ankle flexion and extension were 3/5 and 4/5, respectively. Both right upper and lower limbs had 5/5 power. The deep tendon reflexes were brisk on the left upper and lower limbs, and Babinski sign was positive on the left side. Examination of rest of the neurological system and that of other systems were normal.

His blood investigations were normal. Magnetic resonance imaging (MRI) of the brain with gadolinium contrast showed a 4.0 cm × 5.0 cm × 6.5 cm extra-axial mass lesion in the right mid-third of the parasagittal region [Figure 1a and b]. The lesion was abutting the superior sagittal sinus and extending down along the falx and had multiple hypodense areas within the lesion. It showed weak heterogeneous enhancement with contrast [Figure 1c and d]. There was mild mass effect on the adjacent brain parenchyma.

He underwent a right mid-third parasagittal craniotomy and tumor excision. The specimen was sent for histopathological examination. Following the surgery, his left sided weakness gradually resolved and wound healed. Postoperative contrast-enhanced MRI scan did not show any residual tumor [Figure 1e and f].

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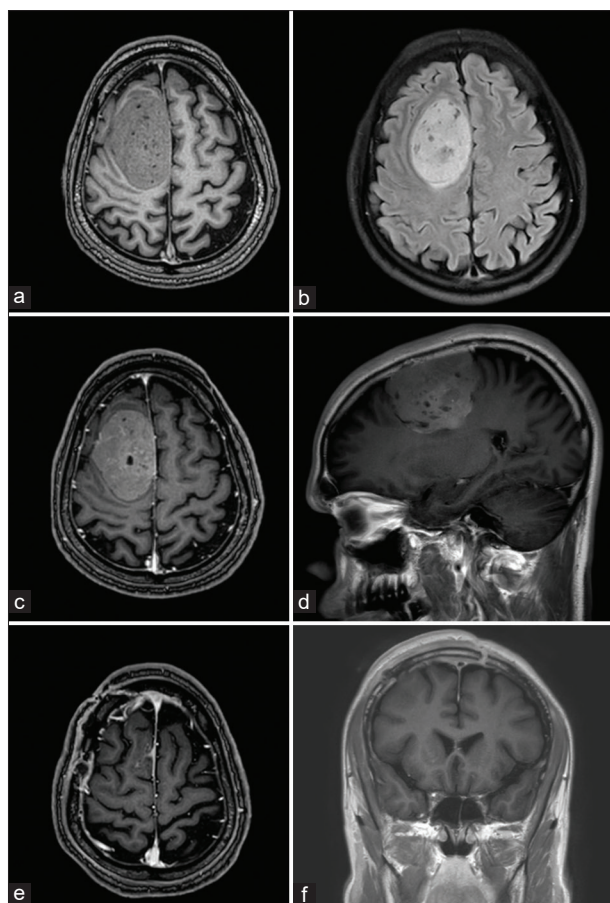


Figure 1: Preoperative axial T1-weighted (a), fluid-attenuated inversion recovery (b), post contrast axial T1-weighted (c) and sagittal T1-weighted (d) magnetic resonance imaging of the brain demonstrated the extra-axial mass lesion in the right mid-third parasagittal region abutting the superior sagittal sinus and extending down along the falx and showed weak contrast enhancement. Postoperative T1-weighted axial (e) and T1-weighted coronal (f) images after IV contrast showed no residual tumor

The specimen was composed of multiple gray-white pieces together measuring 5 cm × 5 cm × 4 cm. Cut section of the larger pieces showed gray-white and yellowish areas. Light microscopy showed a tumor composed of meningothelial cells arranged in sheets and vague whorled pattern. There were multiple aggregates of foamy cells amid the meningothelial cells [Figure 2a and b]. Occasional mitotic figures were noted. There was no pleomorphism, increased cellularity, necrosis, or brain invasion. Immunohistochemistry for epithelial membrane antigen (EMA) showed positivity in the meningothelial and xanthomatous areas [Figure 2c], but CD68 was positive only in the xanthomatous areas [Figure 2d]. Considering these features, a diagnosis of xanthomatous meningioma of the World Health Organization (WHO) Grade I was made.

Discussion

Meningioma is a morphologically heterogeneous tumour. The WHO has classified meningioma based on histology into 15 different types.^[1] Based on biological behavior, they are subclassified into three Grades – I, II, and III. The grade

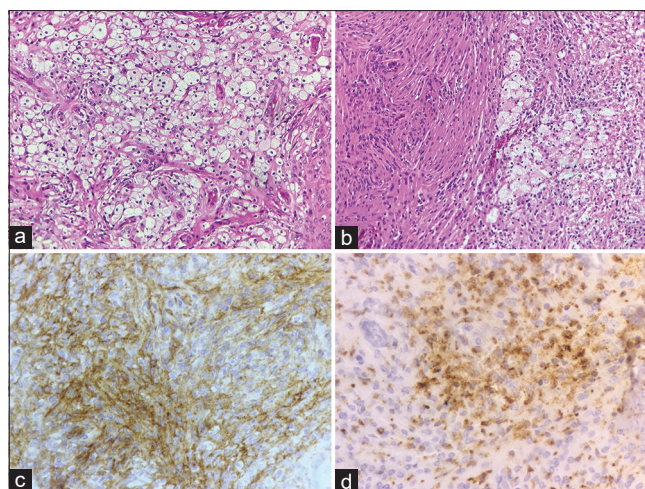


Figure 2: Photomicrograph showing sheets of xanthomatous cells (H and E, ×20) (a), photomicrograph showing meningothelial cells in sheets admixed with xanthomatous cells H and E, ×20 (b), immunohistochemistry stain for EMA showing positivity in the meningothelial and xanthomatous areas. (c) CD68 immunostain showing positivity in the xanthomatous areas (d)

of the tumor has essential bearing on the prognosis of the patient. “Xanthomatous meningioma” was a term coined by Kepes in 1994 and refers to a subclass of metaplastic meningioma.^[2] Metaplastic meningioma is characterized by focal or widespread mesenchymal differentiation in the form of bone, cartilage, fat, or xanthomatous cells. Only a few cases of xanthomatous meningioma have been reported in the literature.

It should be noted that the xanthomatous changes seen in a xanthomatous meningioma are different from the xanthogranulomatous inflammation that may be seen in pyelonephritis and cholecystitis. Xanthogranulomatous inflammation is characterized by exuberant clustering of foamy histiocytes of monocytic derivation with associated chronic inflammatory cell infiltrates.^[3] However, inflammatory changes are not a feature of xanthomatous meningioma. Another differential diagnosis is lipomatous meningioma, which is characterized by meningothelial neoplastic cells with adipocyte-like features. However, these cells which resemble adipocytes exhibit large round eccentric nuclei displaced by large lipid vacuoles resembling mature adipocytes. Another tumor that can lead to diagnostic confusion is microcystic meningioma. This can be differentiated on the basis of numerous cystic spaces filled with edematous fluid forming a cobweb like background by large stellate-shaped meningothelial cells with long cytoplasmic processes, which are highly unusual in xanthomatous meningioma. Finally, glycogen-rich clear cell meningioma, a Grade II tumor, can also appear similar to xanthomatous meningioma and can be differentiated by cells with clear glycogen-rich cytoplasm and prominent blocky stromal and perivascular collagen, which is not a typical feature of the latter.

It is difficult to make a diagnosis of xanthomatous meningioma by radiology. These tumors may appear

hypodense on computed tomography (CT) of the brain.^[4] Hypodensity may sometimes be focal and limited to the lipid part.^[5] The tumors can show variable contrast enhancement leading to an erroneous diagnosis of glioma or metastasis.^[6] A MRI may provide a useful clue to the diagnosis of xanthomatous meningioma when a hypodense mass is demonstrated on CT. On MRI, the tumors may be hyperintense on both T1 and T2-weighted images.^[7] Like the hypodensity on CT, these findings are due to the large amount of lipid in the tumor.

The origin of the xanthomatous cells in these tumors has been debated. Some authors believe that they are meningotheelial macrophages that had possibly migrated to the site of tumor degradation.^[8] This is supported by the fact that xanthomatous meningioma cells express the macrophage immunomarker CD68. However, the expression of CD11c, which is a pan-macrophage marker, has not been documented. It has to be noted that CD68 also stains lysosomes and is not specific to macrophages alone. This has led certain others to speculate that CD68 positivity is due to degenerating tumor cells, which typically contain a large number of lysosomes.^[9] This hypothesis requires further clarification. Another point of view is that the xanthomatous change is merely a degenerative change within the tumor. This view is supported by the expression of EMA in both the xanthomatous and nonxanthomatous components of the tumor.^[10,11] The persistence of the meningotheelial immunohistochemistry profile suggests that xanthomatous change is not true histiocytic metaplasia but rather a degenerative process which occurs in the central portion of the tumor, which is poorly nourished.^[12] Further studies are required to elucidate which of these competing points of view are correct.

Xanthomatous meningioma is one of the rarest variants of metaplastic meningioma with only about 18 cases reported in the literature till date, and this case is the nineteenth one. It was once thought that the meningotheelial-derived tumor cells retained their ability to differentiate, and that is why xanthomatous meningioma is classified under metaplastic meningioma. However, recent studies have identified lipid-filled xanthomatous tumor cells, which were of meningotheelial origin by ultrastructural criteria. These cells showed complex plasmalemmal interdigitations bound by well-formed desmosomes and hemidesmosome-like intercellular specializations.^[2] These features are not typical of macrophages. Moreover, the designation of this tumor as metaplastic has been criticized. Because of these findings, there are those who advocate for a separate class for xanthomatous meningioma, and a new term “lipidized meningioma” or lipomatous meningioma subtype has been proposed.^[13]

Since this is a rare entity, the evidence assessing the clinical behavior of xanthomatous meningioma is weak. However, anecdotal evidence and expert opinion suggest an excellent prognosis.

In conclusion, xanthomatous meningioma is a sporadic type of metaplastic meningioma. Hypodensity on CT scan and hyperintensity on both T1- and T2-weighted MRI images may point toward the diagnosis of xanthomatous meningioma. Definitive diagnosis requires histopathology, and the prognosis in patients with xanthomatous meningioma appears to be excellent.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given consent for images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Approval of ethics committee

The study was approved by the institutional ethics committee.

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Conflicts of interest

There are no conflicts of interest.

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