

**Xanthomatous meningioma; a rare histological variant**Jasim M. A Al-Diab<sup>1</sup>, Hasan H. Almohammed<sup>2</sup>, Sura A. Al-Namil<sup>3</sup>**ABSTRACT**

**Introduction:** Meningiomas are tumors of the central nervous system originating from the meningeothelial tissues, the arachnoid cap cells of the cranium and spinal cord. According to the WHO classification system, grade 1 meningiomas are of nine subtypes. Xanthomatous meningioma is a rare histological variant of meningioma belong to the subtype "Metaplastic meningioma". The Meningioma with a predominantly xanthomatous cellular changes with lipid-laden vacuolated cytoplasm is classified as "xanthomatous meningioma"

**Case report:** A 61 year old patient presented with headache, disorientation and hemiparesis. Magnetic resonance imaging (MRI) shows large right parietal enhancing dural based convexity meningioma. The tumor resected enblock and the histological examination revealed features of classical meningioma with predominant areas of xanthomatous changes. Immunostaining for epithelial membrane antigen and vimentin were positive in both xanthomatous foamy cells and meningeothelial areas. GFAP was negative. CD 68 was positive in few scattered cells in the xanthomatous areas. Ki-67 index was 1% in the highly cellular area. The final histopathological diagnosis was xanthomatous meningioma, WHO grade I.

**Conclusion:** Xanthomatous meningioma is a rare variant of meningioma belongs to the subtype metaplastic meningioma. We present the case because it is an extremely rare worldwide.

**Key words:** Meningioma, xanthomatous, rare histological variant

الاورام السحائية الزانثومية، نوع نسيجي نادر

**المقدمة:** الاورام السحائية هي اورام الجهاز العصبي المركزي الذي ينشأ من الانسجة السحائية العنكبوتية في الجمجمة والحبل الشوكي. ووفقا لنظام تصنيف منظمة الصحة العالمية ، فإن الاورام السحائية ذات الدرجة الاولى تتكون من تسعة انواع فرعية. الورم السحائي الزانثومي هو نوع نادر من الورم السحائي ينتمي الى النوع الفرعي الورم السحائي المتحول الورم السحائي ذات الغالبية العظمى من السيتوبلازم المفرغ بالدهون يسمى "ورم سحائي زانثومي".

**تقرير الحالة:** مريض عمره ٦١ عاما مصاب بالصداع والارتباك وضعف الجانب الايسر من الجسم اظهر التصوير بالرنين المغناطيسي وجود ورم سحائي كبير على الجافية. ازيل الورم كاملا وارسل الى الفحص النسيجي الذي اظهر ملامح ورم السحائي الكلاسيكي مع المناطق السائدة من التغيرات الزانثومية. كان نتائج الاصباغ المناعية لكل من **EMA and vimentin** في كلتا المنطقتين وايجابيا في عدد قليل من الخلايا المتفرقة في المناطق **cd 68**، وسلبيا بالنسبة **GFAP** كان مؤشر الانقسام الخلوي ١% في منطقة الخلوية العالية.

كان التشخيص المرضي النهائي هو ورم سحائي زانثومي.

**الخلاصة:** الورم السحائي الزانثومي هو نوع نادر من الاورام السحائية ينتمي الى الاورام السحائية شبة النمطية. نشر هذه الحالة لأنها نادرة على المستوى العالم.

الكلمات المفتاحية: الاورام السحائية، الزانثومية، نوع نسيجي نادر

**INTRODUCTION**

**X**anthomatous meningiomas are a rare histological variant of meningiomas belong to the subtype "Metaplastic meningioma". The metaplastic meningioma contains different mesenchymal components. They include the cartilaginous meningioma, lipomatous meningioma, melanotic meningioma, myxoid meningioma, osseous meningioma and xanthomatous meningioma. Meningioma with a predominantly

<sup>1</sup>Professor of Pathology, Basrah Medical College

<sup>2</sup>Consultant Neurosurgeon, Al-Sader Teaching Hospital, Basrah

<sup>3</sup>Specialist in Pathology, Al-Sader Teaching Hospital, Basrah

xanthomatous cellular changes with lipid-laden vacuolated cytoplasm is classified as "xanthomatous meningioma".<sup>[1,2]</sup> Xanthomatous meningioma, as a variant of metaplastic meningioma is included in the WHO grade I tumors. According to the World Health Organization (WHO) classification, grade I meningiomas are of nine types; meningothelial, fibrous, transitional, psammomatous, angiomatous, microcystic, secretory, lymphoplasmacyte rich, and metaplastic types(2). The classification of meningiomas has not been revised in the WHO 2016 classification.<sup>[1]</sup> Xanthomatous meningioma needs to be distinguished from the grade 2 clear cell meningioma which is characterized by poorer prognosis with a higher recurrence rate.<sup>[3]</sup>

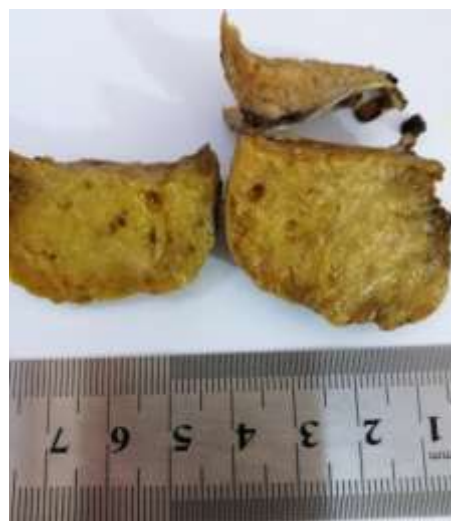
### Case report

Our patient was 61 year old male presented with headache, convulsion and left hemiparesis. Clinical examination reveals cloudy conscious level, disorientation and grade 4+ left hemiparesis. Brain magnetic resonance imaging revealed a large ( 5cm. in maximum dimension) extra axial well defined right dura based parietal strongly enhancing mass (Fig-1).



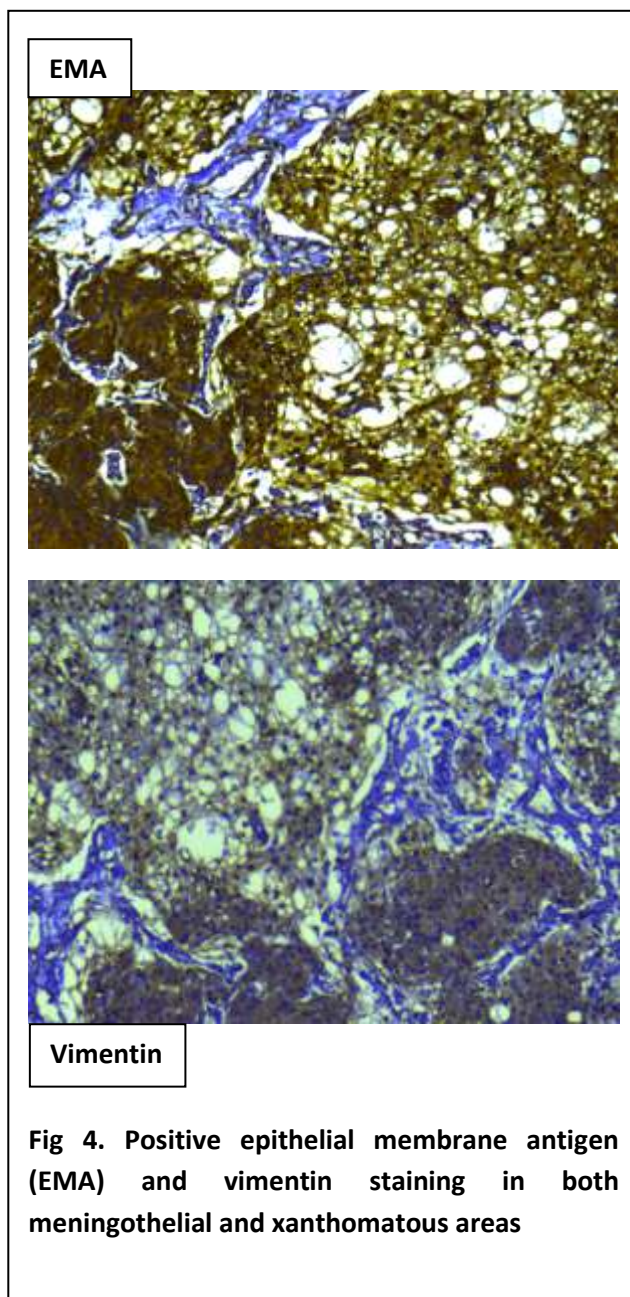
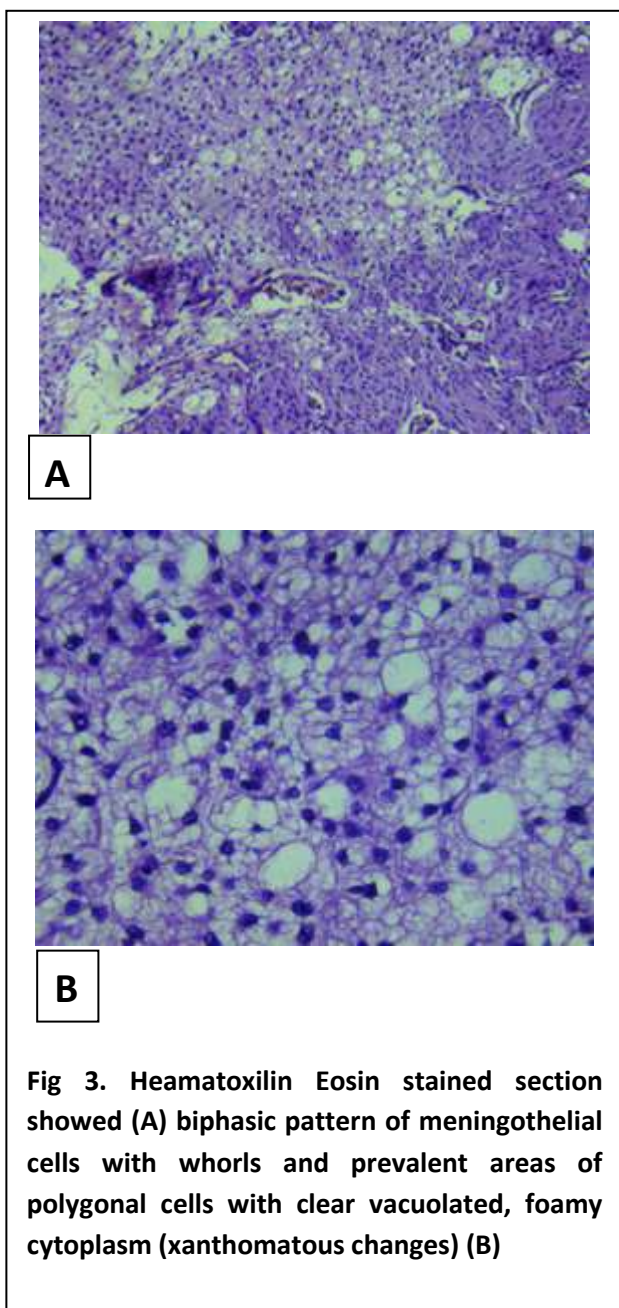
**Fig 1. Brain magnetic resonance imaging revealed a large extra axial well defined right dura based parietal strongly enhancing mass**

The mass was resected enblock and sent for the histopathological laboratory. The macroscopic examination revealed lobulated mass; measures 5x4x4 cm. with smooth outer surface and a piece of dura attached to it. The cut surface was orange yellow in color, firm with small cystic areas (Fig-2).

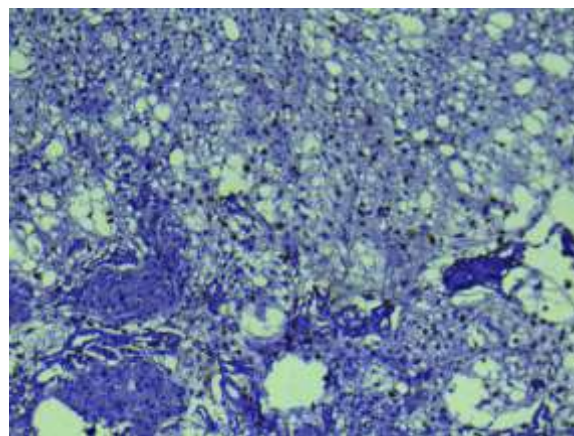


**Fig 2. Cut surface of the tumor is orange yellow in color with small cystic areas**

The microscopic examination revealed diffuse growth pattern of meningothelial cells with many whorls and prevalent areas of polygonal cells with clear vacuolated, foamy cytoplasm (xanthomatous changes). The nuclei are uniform nuclei, atypia, no necrosis and no mitotic activity (Fig-3).



Immunohistochemical staining for Epithelial Membrane Antigen (EMA) and vimentin was positive in both xanthomatous foamy cells and meningothelial areas. GFAP was negative in the xanthomatous and meningothelial areas. Ki-67 index was 1% in the highly cellular area. The histiocytic marker CD68 was positive in few scattered cell in the xanthomatous area (Fig. 4, 5). Taking into consideration the morphological and immunohistochemical findings, the final histopathological diagnosis was xanthomatous meningioma, WHO grade I.



Meningiomas, so named by Harvey Cushing in 1922, are common primary intracranial tumor. They are thought to originate from the arachnoid cap cells (cells forming the outer lining of the arachnoid membrane) of the meninges. Whether the arachnoid cap cells are derived from the neural crest or the mesoderm is itself still controversial<sup>[4]</sup> Grossly most meningiomas are well defined, round or oval, frequently lobulated tumors attached to the dura. The benign meningioma may compress the adjacent brain but not invade it. Most meningiomas are hard to firm in consistency, and on cut surface the typical meningioma is a greyish pink homogenous tumor. Occasionally, yellow flaks representing collection of fat containing cells (xanthoma) are seen.<sup>[7]</sup> Microscopically meningioma are composed of two parts; the first is the basic cellular constituents which are the meningocytic cells arranged, at least in some places, in specific pattern known as whorl. The second component is made up of different forms of tissues which may be absent, minimal or prominent in the tumor.<sup>[10]</sup> The term xanthomatous meningioma should be restricted for a meningioma with a predominantly xanthomatous cellular changes with lipid-laden vacuolated cytoplasm<sup>[1]</sup> Xanthomatous meningioma is a quite rare neoplasm, first described by Kepes in 1994. <sup>[4]</sup> Few cases have been reported worldwide. <sup>[2-6,8]</sup> The most important histological criteria for the diagnosis of xanthomatous meningioma is the presence of xanthomatous tumor cells with clear vacuolated foamy cytoplasm (due to their lipid content) and uniform oval centrally located nuclei.<sup>[6,8]</sup> The xanthomatous cells are morphologically similar to macrophages, but positive staining for epithelial membrane antigen (EMA) and vimentin supports the meningotheial origin of these cells.<sup>[2,6]</sup> In our case the xanthomatous cells are positive for both EMA and vimentin with few scattered foamy cells that are EMA negative but stained positive with the histiocytic marker CD 68. These CD 68 positive foamy cells may be macrophages that

had immigrated to the tumor due to degeneration.<sup>[6]</sup> However, CD68 positivity does not always indicate the histiocytic nature of the cells, it may also reflect the presence of abundant lysosomes in the cytoplasm of the xanthomatous cell. Yet, EMA positivity still the most important stain that support the meningotheial origin of the tumor.<sup>[2,6]</sup> Ishida, et al<sup>[2]</sup> observed adipophilin positivity in both xanthomatous and conventional areas of meningioma. Adipophilin demonstrates lipid droplets in cytoplasm of the tumor cells. However, we didn't use the adipophilin in our case, the foamy appearance was so clear to suggest xanthomatous nature of the tumor cells which are proved to be of meningotheial origin by EMA and vimentin. Xanthomatous meningioma, is classified as grade I meningioma. It needs to be distinguished from the grade II clear cell meningioma which is characterized by poorer prognosis with a higher recurrence rate.<sup>[3]</sup> Another Differential diagnosis of xanthomatous meningioma is the lipomatous meningioma which is characterized by the presence of adipose tissue and it is a very rare subtype of meningioma. The adipocyte-like cells seen in lipomatous meningiomas were reported to be S-100 positive. <sup>[9]</sup> In our current case, we didn't demonstrate adipocyte-like cells in the tumor

## CONCLUSION

Xanthomatous meningioma is a rare variant of meningioma belongs to the subtype "Metaplastic meningioma". Their diagnosis depends on the presence of an appreciated xanthomatous changes in the cytoplasm of the tumor cells together with the presence of conventional meningotheial growth pattern. Awareness should be raised toward this rare variant of meningioma to avoid misdiagnosis.

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