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Journal

International Journal of Surgical Pathology, 26(2)

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Publication Date

2018-04-01

DOI

10.1177/1066896917727100

Peer reviewed

Published in final edited form as:

Int J Surg Pathol. 2018 April ; 26(2): 157–160. doi:10.1177/1066896917727100.

Meningioma With Tyrosine-Rich Crystalloids: A Case Report and Review of the Literature

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Abstract

We report a case of fibrous meningioma with tyrosine-rich crystalloid in the frontal lobe of a middle-aged woman. The patient presented with a history of several years of worsening headaches and blurry vision, which progressed to include syncopal episodes and right-sided weakness. Imaging demonstrated a dural-based extra-axial mass arising from the right orbital roof and extending superiorly along the right frontal convexity causing right-to-left midline shift. The patient underwent a craniotomy and operative resection. Tumor architecture and cytology was similar to that of a Schwannian neoplasm, with spindled cells arranged in a fascicular architecture and displaying focal nuclear palisading. Immunohistochemical stains confirmed a diagnosis of fibrous meningioma. Light microscopy demonstrated extracellular deposits of eosinophilic crystalline material parallel to the spindled tumor cells, reminiscent of “tyrosine-rich” crystals described in salivary gland neoplasms. This is the third meningioma featuring tyrosine-rich crystalloid reported in the literature; we also summarize the previous 2 reports.

Keywords

meningioma; tyrosine; crystalloid

Clinical Case

A 45-year-old woman from Jamaica presented with a history of several years of worsening headaches and blurry vision. Symptoms progressed to include syncopal episodes and right-sided weakness. At physical examination, the patient was afebrile with stable vital signs. Neurological examination was normal.

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Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Imaging Studies

Axial postcontrast computed tomography images demonstrated a large lobulated enhancing mass in the right frontal lobe (Figure 1). Surrounding vasogenic edema and associated mass effect was visible, which resulted in effacement of the frontal horn of the right lateral ventricle and midline shift.

Magnetic resonance imaging (MRI) revealed a large dural based extra-axial mass arising from the right orbital roof and extending superiorly along the right frontal convexity, with no definite intraorbital extension (Figure 2). The mass demonstrated marked heterogeneous enhancement and measured 7.4 × 4.8 cm and showed numerous small cysts. The anterior division of the right middle meningeal artery was asymmetrically enlarged on the right. There was significant mass effect on the anterior falx cerebri with right to left midline shift measuring approximately 11 mm anteriorly and 7 mm at the foramen of Monro. The right lateral ventricle was partially effaced and there was mild entrapment of the left lateral ventricle. Diffusion imaging showed no hyperacute, acute, or early subacute infarction (Figure 2). There was relatively dark signal on apparent diffusion coefficient (ADC) and hypointensity on T2-weighted imaging, which is characteristic of highly dense cellular tissue with a high nuclear-to cytoplasmic ratio, and is seen in meningiomas. Mild to moderate surrounding increased signal on T2/FLAIR-weighted sequences was consistent with edema, although there was no extra-axial collection. No other unusual signaling characteristics of this lesion were noted.

Pathological Findings

The tumor demonstrated several striking histopathologic findings. The tumor architecturally and cytologically mimicked a Schwannian neoplasm, with spindled cells arranged in a fascicular architecture and displaying focal nuclear palisading. Classic features of meningothelial meningioma, such as round to oval nuclei, meningothelial whorls, and psammoma bodies, were lacking. Immunohistochemical stains showed that the neoplasm was positive for epithelial membrane antigen (EMA) and progesterone receptor as well as negative for S100 protein and SOX10, confirming a diagnosis of fibrous meningioma. The MIB-1 proliferation index was approximately 3% to 5%, and 4 to 5 mitotic figures per 10 high-powered fields were identified, leading to the diagnosis of an atypical meningioma (World Health Organization grade II).

The most unusual light microscopic finding was an abundant deposition of extracellular eosinophilic crystalline material (Figure 3). These deposits were seen throughout the tumor but were more numerous in some areas than others. In particular, the tumor showed some foci with “degenerative changes” such as fibrosis, myxoid stromal change, and an increased chronic inflammatory infiltrate, and the deposits appeared more commonly within or adjacent to these foci. An inflammatory response directed at the crystalline material itself was not appreciated. The crystalline deposits were primarily seen arranged in parallel with the fascicles of spindled tumor cells; no clear intracellular crystals were identified. The crystals themselves appeared floret-like with numerous petals arranged around a round to elongated central core. The crystal aggregates varied widely in size, with individual “petals”

measuring 2 to 3 μm in size and large conglomerates measuring up to 20 to 30 μm. These crystals were strongly reminiscent of the “tyrosine-rich” or “tyrosine-like” crystals most commonly described in salivary gland neoplasms, and appeared essentially identical when directly compared against tyrosine-like crystals from a pleomorphic adenoma.[1,2]

Discussion

Here we report abundant extracellular deposition of tyrosine-rich crystalloid in a fibrous meningioma occurring along frontal lobe of a middle-aged woman of African descent. These uncommon crystalline deposits have primarily been described in association with salivary gland neoplasms such as pleomorphic adenoma, myoepithelioma, polymorphous low-grade adenocarcinoma, and adenoid cystic carcinoma.[2] Similar findings have also been documented in fibrous tissue surrounding a squamous cell carcinoma of the larynx.[1]

To our knowledge, this is the third meningioma featuring tyrosine-rich crystalloid to be reported in the literature. The first such tumor was reported was in 1999, and came from a 58-year old African American woman who presented with a 6-month history of headaches.[3] MRI revealed a 5-cm enhancing dura-based mass in the left parietal region, which was composed of uniform spindle cells associated with intercellular collagen and radially arranged clusters of eosinophilic crystals. Tyrosine content was confirmed via a positive Millon reaction. The tumor was diagnosed by light microscopy as a fibrous meningioma.

The second such tumor was reported in 2013, and came from a 54-year old man with a 2-year history of back and right leg pain.[4] MRI of the lumbar spine showed a tumor in the spinal canal at the level of L3-L4, which extended along the right L4 nerve root. Over 7 months, the tumor grew 5 mm to a maximum dimension of 4 cm. The tumor was resected via L3-L4 laminectomy and durotomy. Examination of permanent sections revealed prominent expanses of dense collagenous stromal tissue arranged in broad bands or concentric knots, many of which contained irregularly shaped and sized magenta extracellular deposits. These deposits were similar to those reported by Couce et al,[3] and concluded to also represent tyrosine-rich crystals of the type most commonly seen in salivary gland tumors. However, tyrosine content was not confirmed via histochemistry, that is, reaction with Millon reagent, or via diazotization-coupling reaction. The tumor was diagnosed as a clear cell meningioma based on the clear, glycogen-containing cytoplasm and EMA positivity, despite the unusual presentation as a nondural lumbar spinal tumor.

We note the presence of these rare crystals do not impact the pathologic interpretation of the case. Interestingly, these crystals have been reported exclusively in meningioma and salivary gland tumors from black or African American patients.[2] However, the mechanism by which these tyrosine deposits form has yet to be understood. Pure tyrosine deposits have not been reported to occur in primary tyrosinosis or in experimental excess of tyrosine.[5,6] Finally, this feature is so rare that its prognostic and clinical relevance has not been studied and remains unknown.

Acknowledgments

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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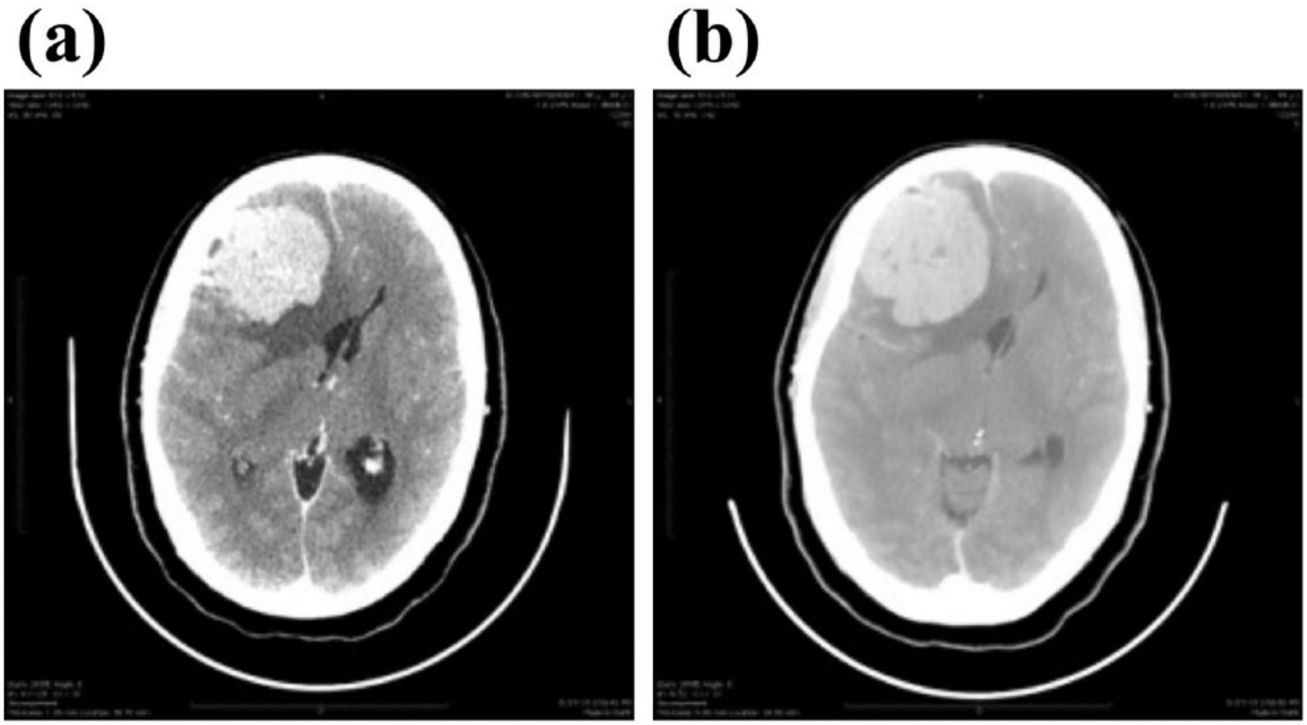

Figure 1.

Axial postcontrast computed tomography scans with (a) soft tissue window and (b) air window demonstrate significant midline shift from meningioma. A large lobulated enhancing mass is along the right frontal lobe, with surrounding vasogenic edema and associated mass effect due to effacement of the frontal horn of the right lateral ventricle and midline shift.

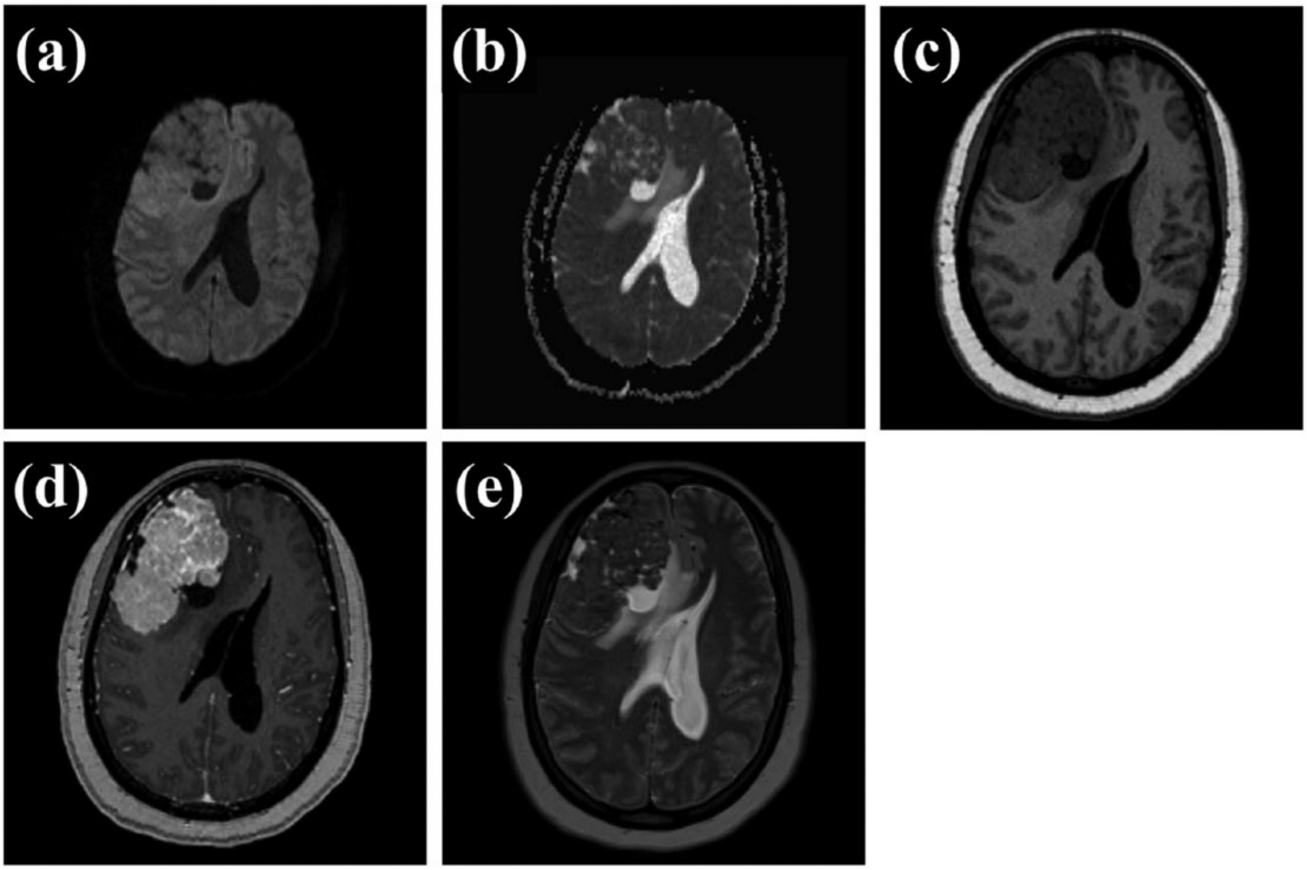

Figure 2.

Axial magnetic resonance imaging (MRI) of the brain performed using a 1.5-T system. (a) Diffusion-weighted with trace weighting, (b) diffusion-weighted with apparent diffusion coefficient map, (c) T1-weighted precontrast, (d) T1-weighted postcontrast, and (e) T2-weighted fast spin echo postcontrast images. There is a large heterogeneously enhancing mass along the right frontal convexity. There are small cysts associated with this mass. There is relatively dark signal on apparent diffusion coefficient (ADC) and hypointensity on T2-weighted image, a characteristic of highly dense cellular tissue with high nuclear-to cytoplasmic ratio such can be seen with meningiomas.

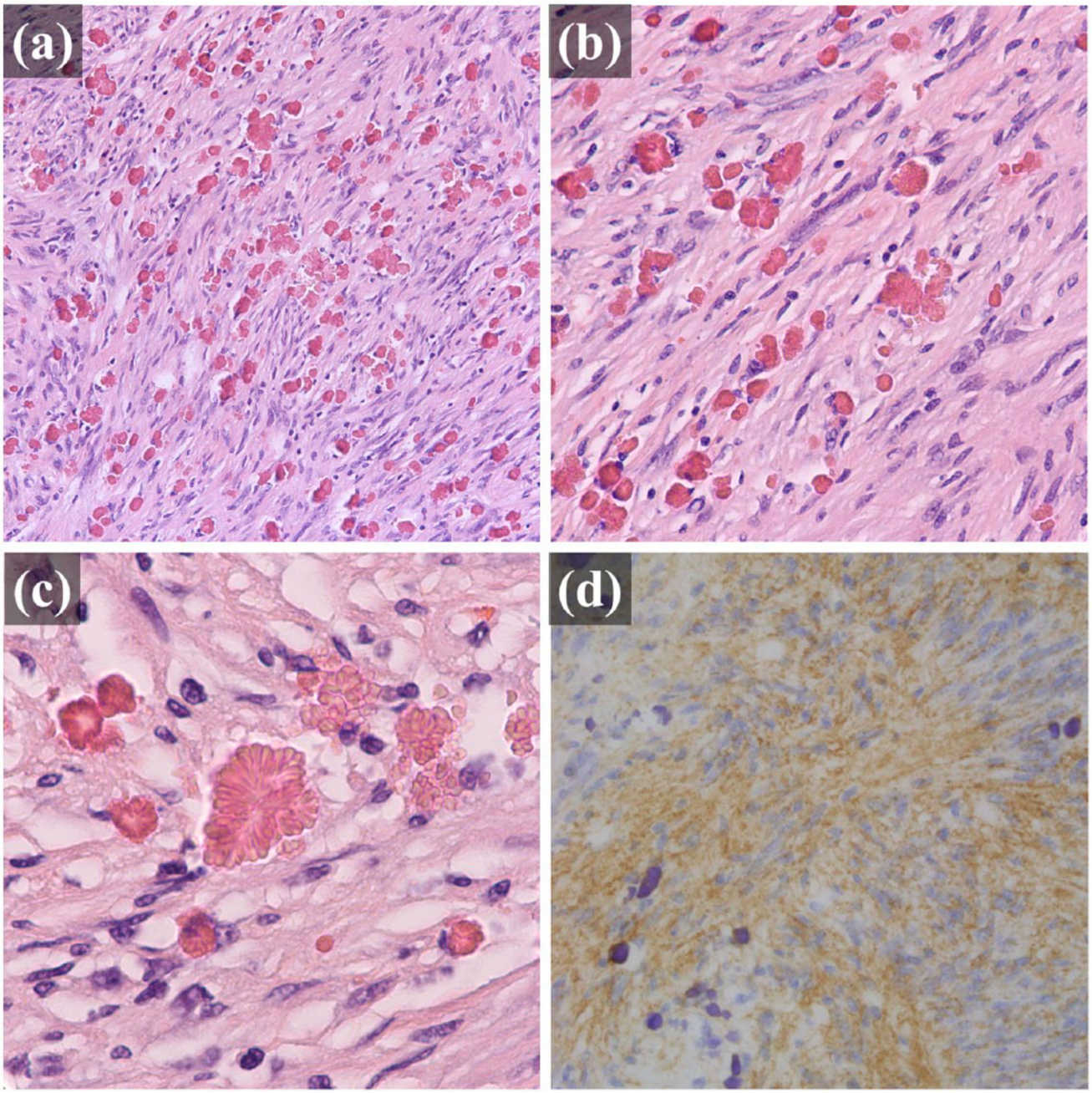

Figure 3.

(a) Hematoxylin and eosin (H&E) 200×: Vague fascicles of spindled cells are seen with numerous eosinophilic deposits. (b) H&E 400×: Eosinophilic crystalloids are present alongside and within tumor cell cytoplasms, (c) H&E 1000×: The crystalloids display a characteristic floret-like shape often described in so-called “tyrosine-rich” crystals, (d) Epithelial membrane antigen (EMA) immunohistochemistry (400×): Membranous and cytoplasmic reactivity as seen in this case is characteristic of meningioma.