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CORRESPONDENCE

Cavernous sinus syndrome as the first presenting sign of metastatic cholangiocarcinoma

Cholangiocarcinoma is poorly treatable and highly lethal adenocarcinoma of the hepatobiliary system. The incidence of brain metastases was 0.15%, 0.47% and 1.4% in three large case series.[1] Cavernous sinus syndrome (CSS) refers to any disease involving the cavernous sinus. It is characterized by ophthalmoplegia, proptosis, chemosis, but also trigeminal sensory loss and Horner's syndrome.[2]

We report the case of a 56-year-old Caucasian woman evaluated due to partial Horner syndrome, periorbital and retroorbital unilateral moderate-to-severe headache involving the left frontal and temporal areas of about 2 weeks duration. She denied trauma, recent illness, numbness, weakness, change in menstrual cycle or recent weight changes. Visual acuity resulted bilaterally normal. Brain magnetic resonance imaging revealed thickening of the cavernous sinus because of the presence of abnormal soft tissue which is isointense on T1, hypointense on T2, with homogeneous enhancement after contrast (Figure 1-3). She was treated with glucocorticoids (intravenous methylprednisolone and oral dexamethasone) considering the hypothesis of Tolosa Hunt syndrome, but without improvement

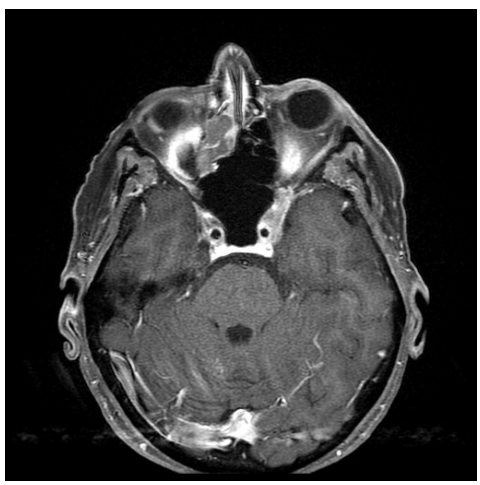

Figure 1

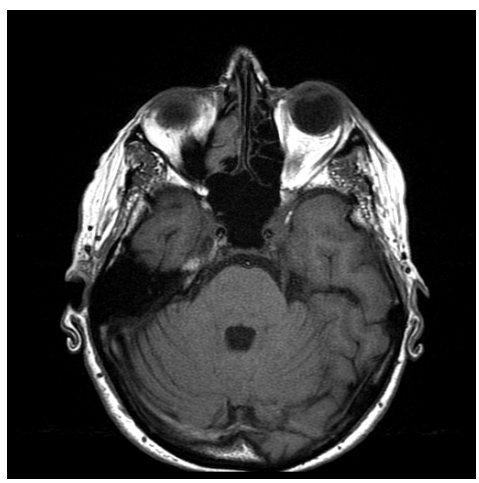

Figure 2

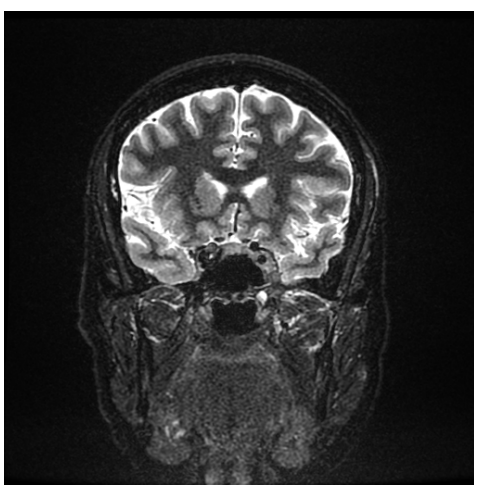

Figure 3

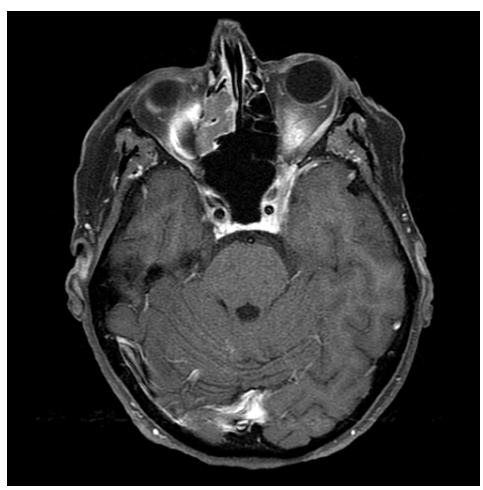

Figure 4

Figures 1-2: axial T1 sequences at baseline showing abnormal soft tissue with thickening of the cavernous sinus
Figure 3: coronal STIR sequence at baseline
Figure 4: axial T1 at follow-up

in pain and neurological examination. After three weeks of rheumatological evaluation, in order to exclude Wegener granulomatosis, chest computer tomography was performed, revealing multiple bilateral round nodule compatible with metastases, while immunological screening resulted normal. Abdomen computer tomography showed adrenal and liver metastases, which were biopsied. This revealed a moderate to poorly differentiated adenocarcinoma, consistent with cholangiocarcinoma, with immunostains positive for cytokeratin (CK)7, CK20, CK19, CDX2. Brain magnetic resonance imaging pattern after one month resulted similar to the former (Figure 4). CA 19.9 resulted >12.000 (normal value <37), while alkaline phosphatase and transaminases resulted all normal, while gamma-glutamyltransferase (68 UI/L) and total bilirubin resulted mildly elevated (1.87 mg/dl). At present time hypofractionated stereotactic radiotherapy (3-30 Gray) has been planned to apply to the brain lesion such as chemotherapy treatment (gemcitabin oxaliplatin- GEMOX).

The previously reported neuro-ophthalmologic presentations of cholangiocarcinoma include one case of a clival mass and sixth cranial nerve palsy, one case of metastasis to the medial rectus muscle and diplopia, two cases of metastasis to the occipital lobe and homonymous hemianopia, and one case of a hypercoagulable state-related stroke and homonymous hemianopia.[3] There have also been two reports of cholangiocarcinoma metastasizing to the orbit presenting with eye pain[4], one report of a combined hepatocellular carcinoma/cholangiocarcinoma metastasizing to the retina and vitreous[5] and an isolated sixth cranial nerve palsy, which was the harbinger of a brain metastatic sellar/suprasellar mass.[1]

This case is unusual due to clinical presentation (CSS) and the difficulty in differential diagnosis with Tolosa Hunt syndrome, due to lack of rapid response to steroid treatment. Biopsy of brain lesion was not performed risk from the lesion site, but we can conclude that the abnormal soft tissue was more likely due to cavernous sinus metastasis rather than an inflammatory lesion.

To our knowledge, this is the first case of cavernous sinus metastasis due to cholangiocarcinoma mimicking Tolosa-Hunt syndrome. We suggest that aggressive metastases of cavernous sinus, including cholangiocarcinoma, should be considered in patients with painful opthalmoplegia without clinical and radiological response with steroid therapy.

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DISCLOSURE

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