



## Case Report

# Suprasellar pleomorphic xanthoastrocytoma: A case report

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Received : 24 July 18

Accepted : 28 November 18

Published : 24 April 19

### DOI

10.25259/SNI-83-2019

### Quick Response Code:



## ABSTRACT

**Background:** Pleomorphic xanthoastrocytoma (PXA) is a rare form of astrocytic neoplasm most commonly found in children and young adults. This neoplasm, which is classified as a Grade II tumor by the World Health Organization classification of tumors of the central nervous system, carries a relatively favorable outcome. It is usually found supratentorially in cortical regions of the cerebral hemispheres, and as such, presenting symptoms are similar to other supratentorial cortical neoplasms; with seizures being a common initial symptom. Due to the rarity of this type of neoplasm, PXA arising elsewhere in the brain is often not included in the initial differential diagnosis.

**Case Description:** This report presents an extremely rare patient with PXA arising in the suprasellar region who presented with progressive peripheral vision loss. Magnetic resonance imaging of the brain demonstrated a heterogeneous suprasellar mass with cystic and enhancing components initially; the most likely differential diagnosis was craniopharyngioma. The patient underwent endoscopic endonasal resection of the tumor. Microscopically, the tumor was consistent with a glial neoplasm with variable morphology. Based on these findings along with further immunohistochemical workup, the patient was diagnosed with a PXA arising in the suprasellar region. At the 1-year follow-up, the patient remained free of recurrence. Although rare PXA originating in other uncommon locations, such as the spinal cord, cerebellum, the ventricular system, and the pineal region have been previously described.

**Conclusion:** Although rare, PXA should be included in the differential diagnosis for solid-cystic tumors arising in the suprasellar region in young adults.

**Keywords:** Brain neoplasm, endonasal endoscopic approach, optic chiasm lesion, pleomorphic xanthoastrocytoma, suprasellar astrocytoma

## INTRODUCTION

Pleomorphic xanthoastrocytoma (PXA), first described by Kepes *et al.*<sup>[8]</sup> in 1979, is a low grade, brain tumor that is thought to arise from the aberrant mitotic activity of sub-pial astrocytes.<sup>[8]</sup> PXA comprises <1% of all brain tumors and is commonly found in young adults and children, with a peak incidence between 10 and 30 years of age.<sup>[6,10]</sup> The vast majority of tumors are found in the supratentorial compartment, with a predilection for the temporal lobes followed by frontal and parietal lobes. The tumors are usually located superficially, often involving the cortex and the leptomeninges, with sparing of the dura. Macroscopically,

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PXAs tend to be well circumscribed with cystic components.<sup>[4]</sup> On imaging, these tumors appear as a solid enhancing nodule frequently accompanied by eccentric peripheral cystic components, surrounding edema and a reactive “dural tail” with leptomeningeal involvement.<sup>[12]</sup> Histologically, these pleomorphic tumors exhibit variable features with spindle cells, multinucleated and fibrillary giant cells, eosinophilic granular bodies, polygonal cells, and lipid-laden xanthomatous astrocytes.<sup>[4,8]</sup> Immunohistochemical analysis demonstrates abundant reticulin, glial fibrillary acidic protein (GFAP) and S100 positivity, and variable staining with neuronal markers including synaptophysin, MAP2, and other neurofilaments.<sup>[10,15]</sup>

Given their predilection for the temporal lobes, many patients with PXA present with progressive headaches or seizures.<sup>[6,12]</sup> However, we report an extremely rare patient who was diagnosed with PXA located in the suprasellar cistern of the third ventricle arising from the optic chiasm.

## CASE REPORT

A 30-year-old man with a 3-year history of progressive loss of peripheral vision in his right eye was referred to our clinic following the discovery of a suprasellar mass on imaging. He denied any headaches, history of seizures, recent changes in weight, breast discharge, heat or cold intolerance, or changes in shoe size or facial features. His only complaint was a progressive visual field defect in the right upper quadrant of the right eye. He had no significant other medical or surgical history, and, apart from a visual field defect of the right eye (quadrantanopia), he had an otherwise unremarkable physical exam including intact cranial nerves III to XII. Endocrine workup was unremarkable. Magnetic resonance imaging (MRI) with and without contrast of his brain revealed a multilobulated complex mass with both cystic and solid components in the suprasellar region measuring 28.7 mm × 34.5 mm × 37.2 mm (AP by TR by CC) [Figure 1]. The radiological differential diagnosis was craniopharyngioma.

## Operation and postoperative course

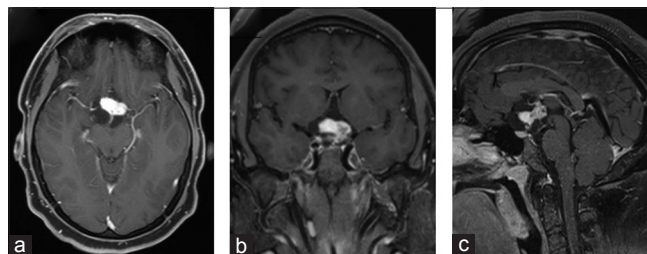
The patient underwent surgery for the resection of the suprasellar tumor through an endoscopic endonasal transsphenoidal approach. The intraoperative visualization of the tumor showed that the mass appeared to be arising from the optic chiasm rather than the Sella. Furthermore, intraoperative resection was complicated by significant adherence of the tumor capsule to local structures, including the carotid arteries. To prevent damage to the surrounding structures, the operation was concluded with an estimated 50–60% of the tumor resected. Intraoperative frozen pathology sections results revealed nonspecific glial reactive tissue. Postoperatively, the patient remained at neurologic baseline except for a new left lower quadrant visual field defect. He did not have any oculomotor nerve palsy. MRI confirmed expected partial resection of the suprasellar mass as well as the decreased

mass effect on surrounding tissues [Figure 2]. The patient’s postoperative course was unremarkable, and he was discharged home. Initially, we considered adjuvant radiotherapy for the tumor that was adherent to the chiasm and carotids; however, the patient’s case was discussed at our hospital neuro-oncology tumor board (neuro-oncologists, pathologists, neurosurgery, and radiation oncology teams) and given the indolent course of the PXA, a decision was made to follow the patient clinically and with serial imaging and if he had a worsening of his symptoms during the follow-up period to perform radiosurgery then.

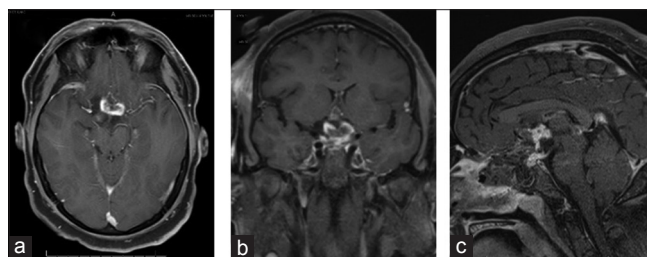
At 1-year follow-up, MRI of his brain revealed the stable size of the known residual with no new areas of enhancement and improvement of the tumor local mass effect. The patient endorsed subjective improvement of his vision, and ophthalmologic follow-up at 1 year revealed partial improvement of his visual fields deficits compared to preoperative evaluation. At 1-year follow-up, the patient was not in any hormonal supplementation.

## Pathological analysis

Microscopically, the tumor was consistent with a glial neoplasm with variable morphology. Some cells were spindled, others were within a mucin-rich matrix, and focal cells with lipidized cytoplasm were present. Eosinophilic granular bodies, lymphocytic infiltrates, and Rosenthal fibers were also noted to be present [Figure 3]. Immunohistochemical workup showed the



**Figure 1:** Preoperative magnetic resonance imaging (MRI) of the brain postcontrast T1-weighted MRI (a) axial view, (b) coronal view, (c) sagittal view reveals multilobulated complex mass with both cystic and solid components again seen in the suprasellar region measuring 28.7 mm × 34.5 mm × 37.2 mm (AP by TR by CC).



**Figure 2:** Postoperative magnetic resonance imaging (MRI) of the brain postcontrast T1-weighted MRI (a) axial view, (b) coronal view, (c) sagittal view reveals partial resection of the previously seen suprasellar mass with decrease mass effect and trace postsurgical hemorrhage.

tumor to be GFAP, vimentin and S-100 positive. Synaptophysin immunohistochemistry showed focal weak positivity, and neurofilament stains showed rare entrapped axons. IDH1 was negative, and trichrome staining showed collagen within the tumor. Ki-67 index was <2%. Additional workup showed diffuse CD34 positivity, positive MAP2 staining, and negative BRAF and V600E staining. Based on these findings,<sup>[4]</sup> a final diagnosis of PXA was made.

## DISCUSSION

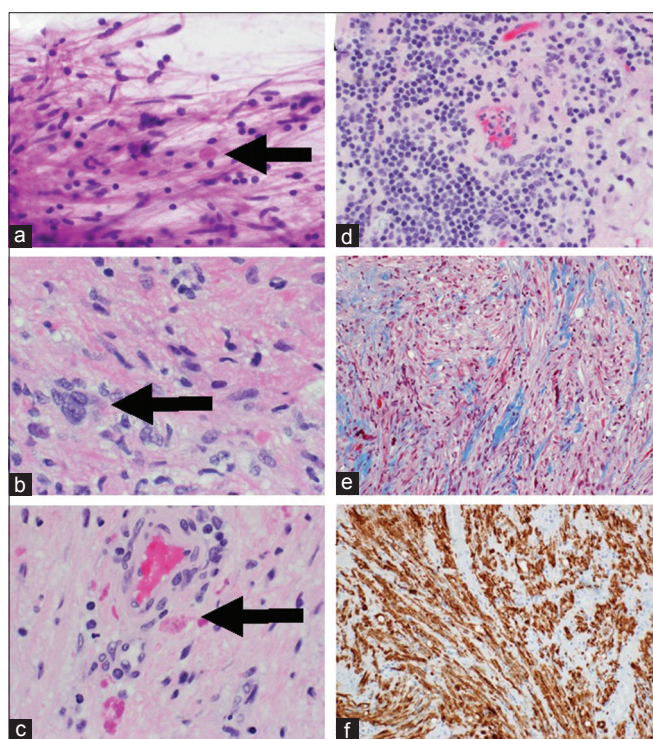
PXA are a rare form of astrocytic tumor that is commonly found in young adults and children.<sup>[4]</sup> Although rare, identification or at least suspicion of, PXA can often be made preoperatively on imaging, given its characteristic location and radiographic morphologic features.<sup>[6,12]</sup> Although pleomorphic in histology this tumor is considered low grade and has a relatively favorable prognosis. Due to PXA's rarity, including it in the preoperative list of differential diagnoses becomes a significant challenge when the tumor does not behave characteristically, especially if the location on imaging

is not the usual. PXA is most commonly found supratentorially, in the cortical regions of the cerebral hemispheres, and typically presents as a cystic mass with a solid, contrast enhancing mural component.<sup>[6,12]</sup> Definitive diagnosis relies on histopathological and immunohistochemistry analysis. The pleomorphic and atypical nature of the neoplastic cells makes an intraoperative diagnosis on frozen sections difficult. As such, PXA can be misdiagnosed as high-grade malignant glioma, and the neurosurgeon may not proceed with complete resection when considering the risks and benefits of the procedure. Therefore, a strong preoperative suspicion of PXA can significantly aid in intraoperative diagnosis and surgical decision-making. Furthermore, multiple reports cite the extent of resection as one of the strongest predictors of recurrence-free survival.<sup>[6,13]</sup> A recent study by Ida *et al.*<sup>[6]</sup> showed that a gross total resection at the initial surgery has a recurrence-free survival at 5 years of 85% compared to 45% for a subtotal resection with stereotactic radiosurgery as adjuvant therapy. These numbers must be taken into consideration for patient and family counseling regarding this type of tumors and in the preoperatively period to prepare for surgery with the goal of a gross total resection.

As our understanding of this tumor changes, new tests are available to further characterize the tumor histologically. The BRAF and V600E mutation, if present, are associated with longer overall survival compared to the tumors that are nonmutant.<sup>[6]</sup> Additional factors to be considered in PXA histology are the presence of multiple mitotic figures. Researchers at the Mayo Clinic initially proposed that PXA that have 5/10 mitotic index per high-power fields should be consider PXA with anaplastic features.<sup>[4]</sup> Recently, they were able to provide further proof that this type of PXA with anaplastic features do harbor a worse prognosis as the overall survival is shorter compared to those without anaplastic features.<sup>[6]</sup> Hence, once the tumor has been resected this additional histologically, and immunohistochemistry tests should be performed to provide additional information for recurrence and overall prognosis for patients with PXA.

As mentioned before PXA is usually a supratentorial tumor with a predisposition to the temporal lobes however, PXA has been sporadically reported to arise in unusual locations including the posterior fossa, spinal cord, and the retina.<sup>[5,11,17]</sup> PXA originating in the suprasellar region is extremely rare and has only been described in six patients in the scientific literature before the case we present here.<sup>[2,7,9,14,16]</sup> In Table 1, we present the summary of the prior 7 cases reported in literature. Review of those reports, in conjunction with this case, seems to suggest that suprasellar PXA tends to occur more frequently in adults rather than children. Furthermore, of the case reports of suprasellar masses [Table 1], the report by Yeh *et al.*<sup>[16]</sup> is the only one in a pediatric patient, and the other cases were in adults from 19 to 78 years old with 4 female and 2 male cases.

On imaging, PXA often presents as an enhancing, solid-cystic mass on MRI, and commonly causes visual disturbances as the presenting clinical symptom due to local mass effect. Interestingly,



**Figure 3:** A squash preparation (a) H and E  $\times 40$  shows spindled cells with fibrillary astrocytic processes and Rosenthal fibers (arrow). Tumor shows focal cellular atypia with occasional "lipidized" cytoplasm (b) H and E;  $\times 60$ ; arrow), readily identifiable Rosenthal fibers (b and c), and numerous eosinophilic granular bodies (c) H and E;  $\times 60$ ; arrow). Perivascular cuffing, chronic lymphocytic infiltrate is present (d) H and E;  $\times 40$ ). Trichrome stain (e)  $\times 20$  highlights the blue-staining connective tissue into which the tumor cells are invading, immunohistochemistry for glial fibrillary acidic protein (f)  $\times 20$  highlights the astrocytic processes of the tumor cells.



**Table 1:** Review of suprasellar PXA case reports available in literature.

Study	Patient demographics age (year)/sex	Clinical presentation	Imaging/operative findings	Treatment	Follow up
Arita 2002 <sup>[2]</sup>	78, Female	Visual disturbance	Enhancing 2.4 cm solid mass	Transsphenoidal surgery, subtotal resection. Gamma-knife (XRT)	No recurrence at 12 months
Yeh 2003 <sup>[16]</sup>	12, Male	Headache, diplopia	Lobulated heterogenous enhancing mass 2.3 cm with cystic component. Optic chiasm displaced, hydrocephalus	Craniotomy for tumor resection	Surgery alone (no data on follow up)
Krossnes 2004 <sup>[9]</sup>	19, Female	Visual disturbance	Enhancing partly cystic 3.2 cm×2.4 cm mass. Mass originating from the chiasm and growing along optic nerves	Craniotomy. subtotal resection with residual along both optic nerves.	No recurrence at 12 years
Vizcaino 2014 <sup>[14]</sup>	39, Female	Visual disturbance	Enhancing mass 1.2 cm×0.9 cm	Surgery (approached not specified) gross total resection	No data
Jiang 2016 <sup>[7]</sup>	29, Male 52, Female	Headaches Dizziness, visual disturbance	3.5 cm×3.0 cm enhancing suprasellar mass with hydrocephalus. 1.4 cm×1.2 cm enhancing suprasellar mass	Surgery (approached not specified) Surgery (approached not specified)	No recurrence at 8 months No recurrence at 12 months

PXA: Pleomorphic xanthoastrocytoma

as with our case, the case by Yeh *et al.*<sup>[16]</sup> was initially thought to be a craniopharyngioma, and the final postsurgical pathology revealed a PXA. Although uncommon, PXA is a potential differential diagnosis for suprasellar masses with cystic and solid components. The suprasellar PXA reported by Krossnes *et al.*<sup>[9]</sup> similarly to our case presented here the tumor originated from the optic chiasm and was growing along the nerves making a total resection high risk for optic nerve damage. As expected, they noticed small areas of enhancement along both optic nerves in their case compatible with residual tumor.<sup>[9]</sup>

As mentioned before by having PXA as a differential diagnosis preoperatively, surgical plan may change to have a more aggressive resection to provide a longer recurrence-free survival. In the larger series of PXAs (74 cases), Ida *et al.*<sup>[6]</sup> reported a 64.6% of recurrence free at 5 years for both PXA and PXA with anaplastic features. Although in general PXAs have a favorable outcome, recurrence and disease progression are seen in 15–20% of cases, requiring long-term follow-up especially in cases with a subtotal resection. In our case at 12 months, the patient was free of recurrence, and the known residual tumor was stable in size.

At present, there is not a definite guide to treat PXA that did not have a gross total resection during surgery. As mentioned before in general for all PXA, a gross total resection is one of the most important factors for recurrence-free survival.<sup>[6]</sup> For subtotal resections only, a handful of report exists for adjuvant therapy and most of them will start once the patient has had a disease recurrence. There is no current guideline for coadjuvant therapy at the time of surgery for subtotal resection, and most of the treatments are extrapolated from similar tumors histology and grade in the same location. Of the suprasellar PXAs cases available in the scientific literature only the case by Arita *et al.*<sup>[2]</sup> had a subtotal resection

and coadjuvant stereotactic radiosurgery. In our case given the overall indolent course of the tumor and the improvement of the patient's visual symptoms, we elected to monitor him clinically and with serial MRI of his brain and proceed with adjuvant therapy if new symptoms develop or his vision worsens. Ida *et al.*<sup>[6]</sup> proposed that for a tumor that displays anaplastic characteristics adjuvant therapy should be considered right away due to their high frequency of recurrence. There are some examples of good response in anaplastic PXA to adjuvant therapy<sup>[1,3]</sup> On the other hand, if gross total resection is achieved the literature supports that there is no need for immediate coadjuvant therapies.

## CONCLUSION

PXA are rare neoplasms, but they should be considered in the differential diagnosis of suprasellar solid-cystic tumors, especially those found in children and young adults. Gross total resection provides the best overall recurrence free for these tumors and should be the initial goal of surgery: coadjuvant therapy can be considered for tumors with anaplastic features.

## Declaration of patient consent

Patient consent was not obtained as the patient is covered under the Institutional Review Boards protocol.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

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**How to cite this article:** Telemi E, Martirosyan NL, Avila MJ, Lukefahr AL, Le C, Lemole GM. Suprasellar pleomorphic xanthoastrocytoma: A case report. *Surg Neurol Int* 2019;10:72.