Toothy Craniopharyngioma: A literature review and case report of Craniopharyngioma with extensive odontogenic differentiation and tooth formation

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Introduction:

A young child presenting with a craniopharyngioma (pituitary adamantinoma) demonstrated the rare phenomenon of pronounced odontogenic elements / tooth formation on imaging. The origin of the craniopharyngioma from primitive stomatodeum theoretically facilitates odontogenesis (1). Theories of the genesis of this tumour include remnants of the craniopharyngeal duct, squamous metaplasia of anterior pituitary cells and misplaced tissue of the embryonic enamel organ (2). A literature review documents six cases of craniopharyngioma with tooth formation although none appear to be as extensive as the case presented. Craniopharyngioma should be added to teratoma in the differential diagnosis of suprasellar odontogenic elements demonstrated by imaging studies.

Case report:

A 2 year 9 month old girl presented with headache and irritability after sustaining head trauma during a fall. In addition the parents noticed the child's poor vision, delayed milestones and more recent progressive left pupillary dilation and left-sided ptosis. An initial CT scan of the brain demonstrated a large well circumscribed tumour centred on the suprasellar region but also involving and expanding the pituitary fossa. The striking feature of the mass was the presence of innumerable discreet high density elements resembling teeth (Fig 1 a, b and c and Fig 2 a, b). It was difficult to fully assess the intervening soft tissue component due to artefact caused by the densely calcified elements. There was significant mass effect on the structures of the middle fossa with obstructive hydrocephalus. The age of the child and the imaging characteristics led to a preliminary diagnosis of teratoma.

The patient was referred for an emergency shunt procedure. A post-shunt plain film examination of the skull was performed for shunt evaluation. This demonstrated the surgical shunt, splaying of the cranial sutures and a copper beaten / lacunar calvarium reflecting chronic raised intracranial pressure. The odontogenic elements of the tumour were also clearly demonstrated as well as expansion of the pituitary fossa (Fig 3).

The patient also had an MRI of the brain for pre operative planning (Fig 4 a, b and c).   The mass was clearly delineated with internal focal low signal intensity foci on T1W, T2W and GRE sequences in keeping with the calcified odontogenic elements. The intervening soft tissue component was iso to low signal on T1W and high signal on T2W and FLAIR. Post administration of gadolinium the soft tissue component demonstrated heterogeneous enhancement. The adjacent brain parenchyma showed minimal oedema. The left lateral ventricle was decompressed by the previously inserted shunt; however the right lateral ventricular system remained obstructed at the level of the interventricular foramen. The pituitary gland and hypothalamic structures could not be clearly discerned from the tumour mass.

At surgery the tumour was approached from the right fronto-temporal region with opening of the Sylvian fissure. The tumour mass was exposed and the optic nerves/chiasm and circle of Willis were identified.   In order to remove the tumour, each tooth had to be individually resected. There were no intra operative complications and the patient was extubated post operatively with no new neurological signs.

During the first two post operative days the child seemed to be making a good recovery. However on the third day the patient developed diabetes insipidus. Cerebral oedema and neurogenic pulmonary oedema followed and the patient unfortunately died due to hypothalamic crisis.

Histological examination of the tumour revealed an adamantinomatous craniopharyngioma with pronounced odontogenic elements.

Microscopic examination confirmed the characteristic histology of craniopharyngioma (adamantinomatous-type), with a small amount of adjacent premorbid adenohypophysis (Fig 5). Additionally, odontogenic rests and pronounced odontogenic differentiation was present in the surrounding fibrous stroma which revealed multiple well-formed teeth (Fig 6 and 7). The latter demonstrated an organoid appearance with central dental papilla, adjacent odontoblastic layer, dentine, enamel, external ameloblasts and peripheral enamel organ. No definite endo-/meso-/ectodermal teratomatous elements were identified.

Discussion:

Craniopharyngioma is considered the commonest intracranial tumour of childhood and adolescence accounting for 8.3% of intracranial tumours and 14.5% of all supratentorial tumours. These tumours grow as a pseudoencapsulated mass, usually in a suprasellar location and less commonly are intrasellar. Suprasellar tumours are usually both solid and cystic. Although they often destroy the pituitary, they may also grow toward the third ventricle, optic chiasm and tract, pons and thalamus. Through continued growth and pressure, the sella turcica is destroyed, as is the pituitary (2).

Craniopharyngiomas are classically divided into two subtypes: adamantinomatous and papillary. The adamantinomatous subtype is most commonly found in the paediatric age group and usually presents as a complex mixed cystic and solid mass which characteristically contains flecks of calcification (3). On MR imaging the cystic component is usually hyperintense on T1W due to the high protein content of the cysts while the solid component is usually of heterogeneous signal on T1W and T2W images. The solid component enhances heterogeneously. Gradient echo sequences are useful for the presence of calcification confirmed by demonstrating a 'blooming artefact'. The papillary subtype is more common in the adult population (>50years) and is more commonly solid and less likely to contain calcification than the adamantinomatous subtype (3). Our case did not demonstrate typical features of craniopharyngioma.

Many theories have been postulated regarding the origin of these tumours and include:

1. Epithelial rests and craniopharyngioma arising from a portion of an incompletely obliterated craniopharyngeal duct (4).
2. A metaplastic process where squamous cells might arise from other cell types.
3. The tumour could arise by an inrolling of dental elements into the tuberal process of the developing pituitary (5). Whether the tumour arises from embryonic residues or metaplasia, the similarity to the adamantinoma of the jaw is valid only if the embryonic ductal epithelium remains competent to differentiate in this direction (6).

The differential diagnosis includes epidermoid, dermoid and teratomas (2).

Conclusion:

The diagnosis of craniopharyngioma is usually made when a tumour exhibits the classic imaging features. The presence of tooth like structures within a tumour primarily raises suspicion for teratoma. The intimate embryological origin of tumours arising from Rathkes Pouch should be kept in mind when diagnosing tumours in the sellar / suprasellar region.

References

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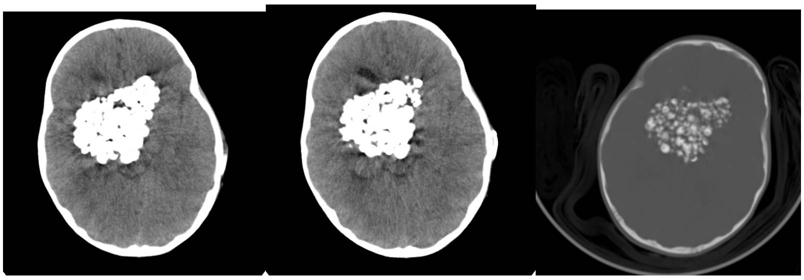
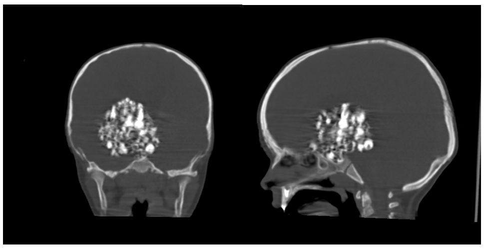
Images

Figure 1a, b, and c: Axial Pre-contrast CT scan reveals a large dense lesion. Bone algorithm reveals the densely calcified elements



a b c

Figure 2 a and b: Coronal and sagittal reconstructions demonstrate the supra-sellar location of the lesion



a b

Figure 3: Lateral skull radiograph reveals the position of the VP shunt, the splaying of the sutures and the “copper beaten skull” appearance

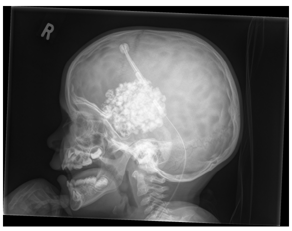


Figure 4 a, b and c: Axial T2 and T1 post gadolinium coronal and sagittal images demonstrates the ‘blooming’ artefact of the dense calcifications as well as the supra-sella location of the mass

a b c

Figure 5: Conventional adamantinomatous-type craniopharyngioma component (arrows) in relation to premorbid adenohypophyseal tissue (asterisks) (H & E, original magnification x 100)

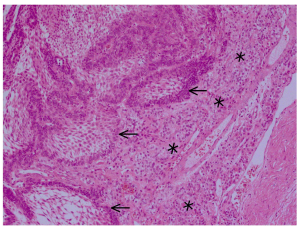


Figure 6: Primitive odontogenic mesenchyme and epithelium with rudimentary tooth formation (H & E, original magnification x 100)

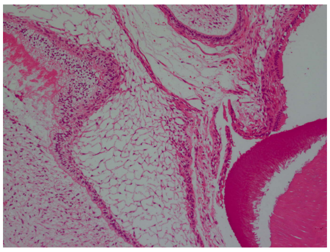


Figure 7: One of multiple tooth structures demonstrating readily identifiable dental pulp, dentine, enamel and surrounding odontogenic epithelium (H & E, original magnification x 40).