Primary Craniopharyngioma in the Ethmoid Sinus: An Unusual Presentation

Joyce HO, Eugene WONG, Murray SMITH, Niranjan SRITHARAN
Department of Otolaryngology Head and Neck Surgery, Westmead Hospital, Sydney, Australia

Background
- Craniopharyngiomas are benign but aggressive epithelial tumours that arise from Rathke’s pouch.\(^1\)
- Usually found in the sellar and suprasellar region.
- Primary isolated ethmoid craniopharyngioma is extremely rare and there have only been two other case reports of this in the literature.
- We describe the third case of this rare condition, and the first in the adult population.

Case Presentation
- 31-year-old female presented with long history of nasal obstruction.
- Examination confirmed a right-sided deviated nasal septum, bilateral inferior turbinate hypertrophy, and a left nasal cavity lesion.
- CT of paranasal sinuses demonstrated a lobulated partly ossific density arising from the left ethmoid sinuses, measuring 17 x 13 mm, with minor soft tissue component.
- Patient underwent a septoplasty, inferior turbinoplasties, and left endoscopic sinus surgery with removal of the mass.
- Intra-operative findings included a bony mass filling the left ostiomeatal complex, and a stenosed left maxillary ostium.
- Histopathology of the left anterior ethmoid mass demonstrated features of an adamantinomatous craniopharyngioma.
  - Large vaguely polyoid portion of respiratory type mucosa with focal squamous metaplasia.
  - Underlying fibrovascular connective tissue shows presence of vital cortico-cancellous bone with intervening loose fibrovascular marrow.
  - Intimately associated with the bone is a multifocal tumour comprising ameloblastoma-like epithelium composed of follicles and a plexiform arrangement of the cells.
  - Follicles exhibit central stellate reticulum-like epithelium with peripherally palisaded columnar cells.
  - At 5 weeks follow-up, patient was asymptomatic.

Discussion
- Craniopharyngiomas originate from the squamous remnants of an incompletely involuted craniopharyngeal duct (which also develops from Rathke’s cyst).\(^1\)
- Radiologically, often heterogeneous due to presence of cystic and calcific components.
- Histopathological confirmation is necessary for diagnosis.
- Two main subtypes of craniopharyngiomas: adamantinomatous and papillary.
- Radio-resistant tumours – therefore, surgical intervention is the treatment of choice.
  - Complete surgical resection is the aim, in order to reduce the risk of local recurrence.
- Two previous cases of primary ethmoidal craniopharyngiomas have been reported.
  - 17-year-old male with left massive epistaxis following biopsy of an ipsilateral ethmoidal lesion, which was confirmed to be an adamantinomatous type craniopharyngioma.\(^2\)
  - 7-year-old boy male with several month history of intermittent epistaxis. Soft tissue density over left ethmoid sinus confirmed to be a craniopharyngioma.\(^3\)

Conclusion
- Craniopharyngiomas are rarely found outside of the sellar and suprasellar regions.
- Primary extracranial craniopharyngioma should be considered as a differential diagnosis when assessing sinonasal tumours.
- Surgical resection of primary ethmoidal craniopharyngiomas can be performed safely using endoscopic techniques.

References