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Craniopharyngioma in a 7-Year-Old Child

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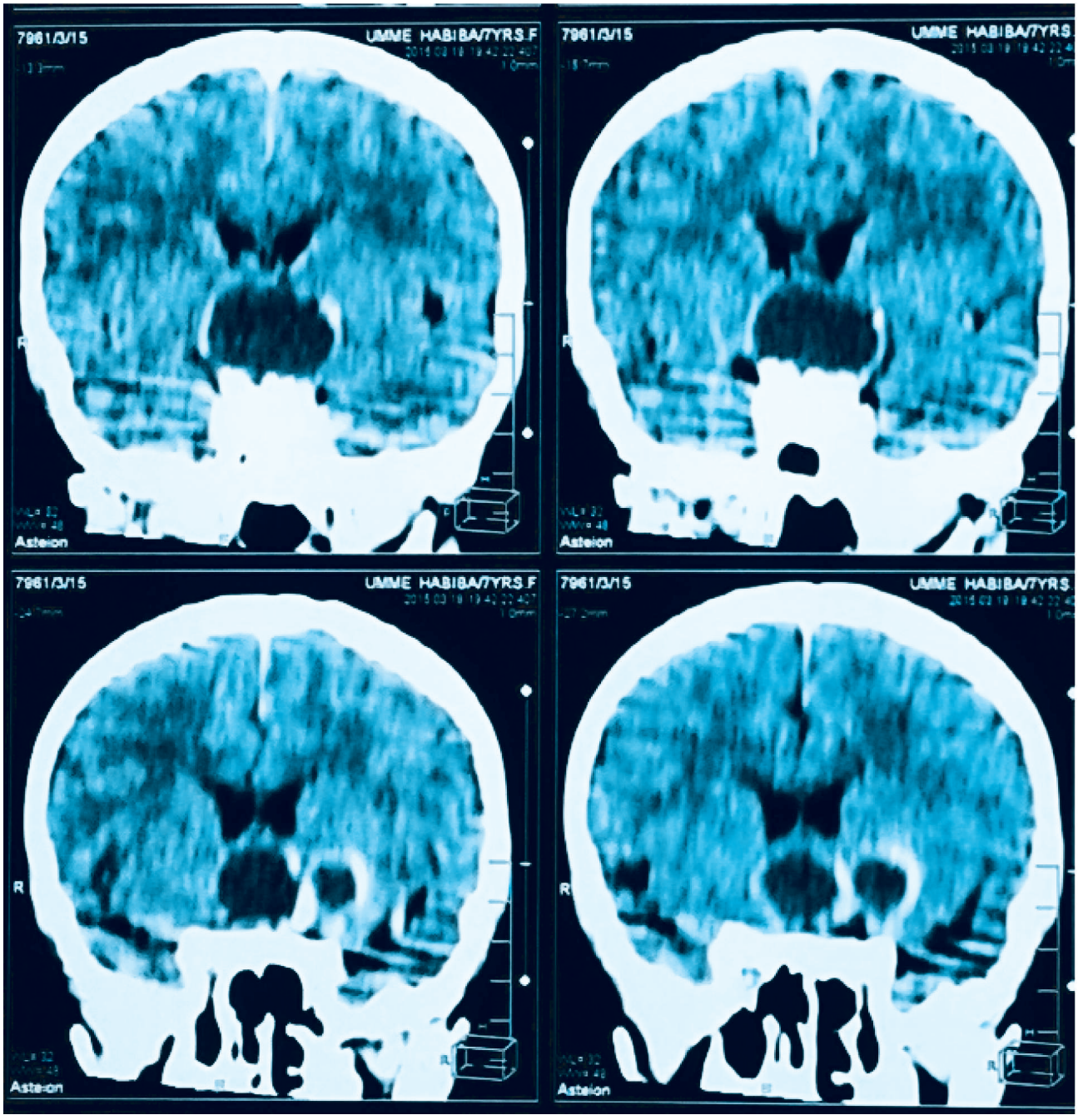

Fig 1. A loculated multicystic mass in the midline suprasellar region with heterogenous soft tissue nodules (coronal section)

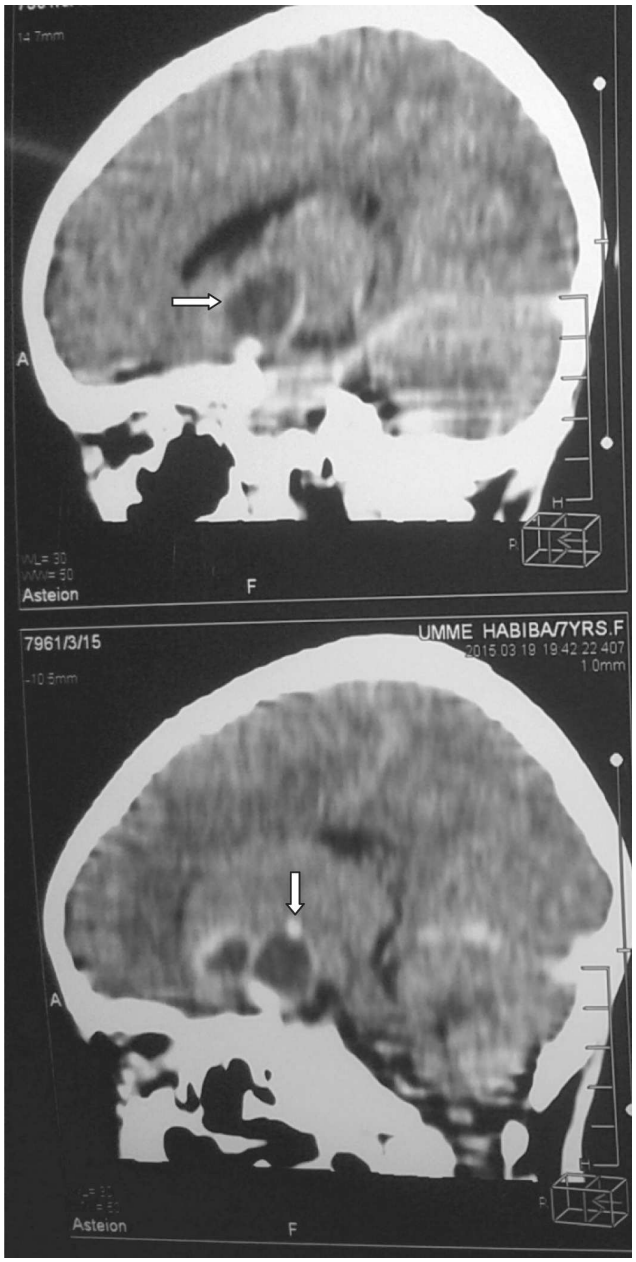

Fig 2. A loculated multicystic mass in the midline suprasellar region with heterogenous soft tissue nodules (sagittal section)

A 7-year-old girl was admitted in the Department of Paediatrics with history of repeated vomiting for 15 days and occasional fever and headache for same duration. On examination she was found conscious but lethargic. She was afebrile, pulse was 92/min and BP was 90/50 mm of Hg. On nervous system examination cranial nerves, motor and sensory systems were intact. There were no signs of meningeal irritation. Complete blood count of this patient showed normal findings. Serum electrolyte showed hypernatraemia and hypokalaemia. There was no papilloedema. CSF study was normal. CT scan of brain showed loculated multicystic mass of about 43 × 31 mm in the midline suprasellar region with heterogenous soft tissue nodule. There were multiple small calcifications in the wall. These features are suggestive of craniopharyngioma. We treated the patient conservatively and referred her to the department of Neurosurgery for further management.

Craniopharyngioma is a tumour of childhood accounting for 7–10% of all childhood tumours and 50% of all sellar/parasellar tumours.[1] The incidence of newly diagnosed craniopharyngiomas ranges from 0.13 to 2 per 100,000 population per year, with a point prevalence of 1 to 3 per 100,000 population with the peak incidence at 5–14 years.[2,3] Craniopharyngiomas are thought to arise from epithelial remnants of the craniopharyngeal duct or Rathke's pouch (adamantinomatous type) or from metaplasia of squamous epithelial cell rests that are remnants of the part of the stomadeum that contributed to the buccal mucosa (squamous papillary type). These tumours are heterogenous in nature displaying both solid and cystic components occurring within suprasellar region. They are minimally invasive, adhere to adjacent brain parenchyma.

The diagnosis of a patient with a craniopharyngioma is based on clinical (neurological and endocrine symptoms) and radiological (a calcified solid/cystic mass) findings, and is then confirmed by characteristic histological findings.

The onset of symptoms is normally insidious with most patients at diagnosis having neurological (headache, vomiting, visual disturbances) and endocrine (growth retardation, delayed puberty) dysfunctions. The classical appearance of a craniopharyngioma is of a sellar/parasellar partly solid and partly cystic calcified mass lesion.[4] The calcification is best delineated on computerised tomography (CT) (Fig 1, 2). Magnetic resonance imaging (MRI) with and without contrast will, however, more accurately delineate the extent of the tumour and in particular, its involvement with the hypothalamus. It is the investigation of choice to plan the surgical approach.

There are two main management pathways with regards to the treatment of the tumour. The first involves attempted gross total resection of the tumour[5,6], the second approach is for more limited surgery, aimed at debulking the tumour to reduce the mass effect on the optic pathways and/or to re-establish the cerebrospinal fluid (CSF) pathways followed by radiotherapy.[7] There is no place for systemic chemotherapy; now-a-days-immunological therapy is being used.[8] The overall five-year survival is 80% but the survival is better in children (5 year survival rate is 85%) than in older adults (5 year survival is 40%).[9]

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