In April 2007, a 21-year-old woman presented with secondary amenorrhoea for 5 years. Her past medical history included bilateral valgus heels with difficulty in walking as a toddler, fractured nose and epistaxis with nasal septum deviation (seen in ENT clinic in 1998). She had a family history of migraine and was referred to the neurology clinic for throbbing headaches in the right frontal region radiating to the occipital region and a few episodes of blurred vision with no focal neurological deficit; migraine was diagnosed and she was prescribed Nortriptyline in 2002. She was discharged from the neurology clinic in May 2004. She was again seen in ENT clinic in 2004 for right-sided tinnitus; her eardrums, external auditory meati, pure tone audiometry and tympanograms were normal bilaterally, and hence it was presumed that her tinnitus was most likely associated with migraine and she was discharged from the ENT clinic.

She was seen in the gynaecology clinic in 2006 for 5-year history of secondary amenorrhoea with presumed diagnosis of polycystic ovarian syndrome and then referred to us in the Joint Gynae-Endocrine clinic in 2007. Systemic examination was normal; body mass index was raised at 27. Visual field testing showed generalized reduction in sensitivity affecting both eyes, more in the lower visual fields with minimal patchy areas of reduced sensitivity. Her hormonal profile revealed normal luteinising hormone, follicle stimulating hormone and oestradiol; raised prolactin (738, normal range (NR) <496 μu/l), negative macroprolactin and cortisol (903; NR 170–540 nmol/l); however, 24-h urine cortisol was normal. Rest of biochemistry was normal and chromosomal analysis showed normal female karyotype (46XX). Magnetic resonance imaging (MRI) of the brain revealed a large cystic lesion in the hypothalamic area causing deformity of the third ventricle, extending from the floor of the lateral ventricle to the suprasellar cistern and in contact with the left half of the pituitary gland, associated with early hydrocephalus, consistent with craniopharyngioma (Figure 1). Neuroendoscopic cystostomy for a cystic suprasellar craniopharyngioma was planned and drainage of the cystic hypothalamic lesion was performed in March 2008; the procedure was not completed due to intraoperative bleeding. There was minimal cerebrospinal fluid (CSF) leak post-operatively. Repeated MRI in May 2008 showed significant reduction in the size of the suprasellar cyst after drainage. Recent pituitary MRI has shown a slight increase in the anteroposterior diameter of the lesion; however, the maximum transverse diameter has remained unchanged. One year post-operatively, she had slightly raised prolactin (983 μu/l) and low gonadotrophins with slightly low oestradiol; she was started on oral contraceptive pill. Short synacthen test, thyroid function and growth hormone are normal. Her headaches gradually improved but never completely resolved. She
Discussion

Headache is a common symptom referred to the medical or neurology clinics, associated with other non-specific symptoms. This article describes a case of a young girl who had headaches, blurred vision, tinnitus and secondary amenorrhoea for many years was seen in various speciality clinics before the diagnosis of craniopharyngioma was made.

Craniopharyngioma are slow-growing benign cystic tumours of the sellar and parasellar area with an overall incidence of 0.2–2 new cases per million population per year; 30–50% of all cases occur in childhood. Clinical features include visual disturbances, endocrine deficiencies, signs due to raised intracranial pressure, as in our patient, unilateral hearing loss, obesity, pubertal delay in teenagers and cognitive impairment in adults. Spontaneous rupture of craniopharyngioma is a rare phenomenon but has been described. These are associated with significant long-term pituitary dysfunction. Malignant transformation following radiotherapy has been described. Ideal surgical treatment remains a major challenge for neurosurgeons as these tumours grow in the deep-seated hypothalamic area that is paramount for vegetative, emotional and endocrine function and good quality for life; it involves balancing adequate reduction in tumour volume and prevention of recurrence while minimizing damage to delicate surrounding structures. Surgical excision followed by external beam irradiation is the main treatment option. Extended endoscopic endonasal approach seems to provide a valid alternative to transcranial approach. Total removal in 60–70% of patients can be achieved, followed by radiotherapy; recurrence is a problem in 15% of patients with total removal. New hypopituitarism after surgery occurs in approximately one-third of the patients. Gamma-knife radiosurgery may provide a favourable benefit as shrinkage of tumour was achieved in 91% of patients in one study, with no visual or neuro-endocrine complications.

Conflict of interest: None declared.

References