CENTRAL PRECOCIOUS PUBERTY AS THE INITIAL MANIFESTATION OF A PILOCYTIC ASTROCYTOMA WITH LEPTOMENINGEAL DISSEMINATION SIMULATING A HYPOTHALAMIC DYSGERMINOMA

B. Galo, D. Yeste Fernández, A. Campos, M. Clemente León, A. Llort (*), A. Carrascosa Lezcano.
Department of Paediatric Endocrinology (*), Department of Paediatric Oncohaematology. Vall d'Hebron University Hospital, Barcelona.

INTRODUCTION

Pilocytic astrocytoma (PA) is the most common type of central nervous system (CNS) astrocytoma in children. Clinical manifestations depend on its location and size and initial symptoms are usually related to neurological deficits or signs and symptoms of intracranial hypertension. Involvement of the hypothalamic area, pituitary infundibulum and leptomeningeal spread are exceptional.

CASE REPORT

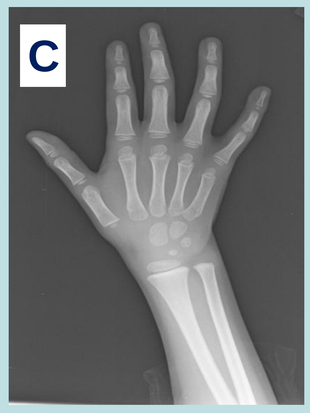
A 4–year-old boy was referred for study of pubic hair of 3 months' evolution with no other accompanying symptoms. Family and personal history were normal.

Physical examination
Weight: 17.5 kg (-0.7 SD)
Height: 116.8 cm (+2.4 SD)
Tanner: G1P2
Testicular volume: 5-4 ml

Fig. A and B

Bone age
4 years, 6 months

Fig. C



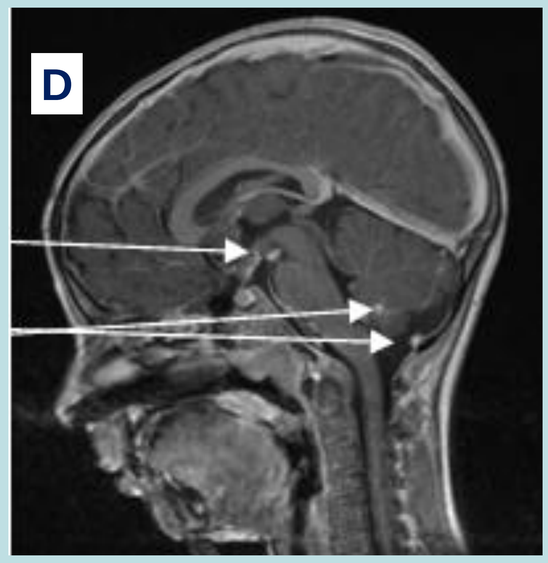
Basal hormonal study
Adrenal androgens and thyroid hormones: normal. LHRH test: basal LH: 1.4 IU/L; peak LH: 23.0 IU/L and basal FSH: 2.3 IU/L, peak: 4.4 IU/L, testosterone: 65.9 ng/dl, prolactin: 6.0 ng/ml, IGF -1: 139.2 ng/ml, ACTH: 52.4 pg/ml, basal cortisol: 15.0 mcg/dl and urine osmolarity after a dry dinner: 875 mOsm/Kg. Study of serum tumour markers (alpha-fetoprotein and hCG beta subunit): negative.

Brain MRI
Hypothalamic thickening with left predominance, consistent with germ cell tumour. Nodular images of supra- and infratentorial leptomeningeal dissemination.

Fig. D

Tumour infiltration

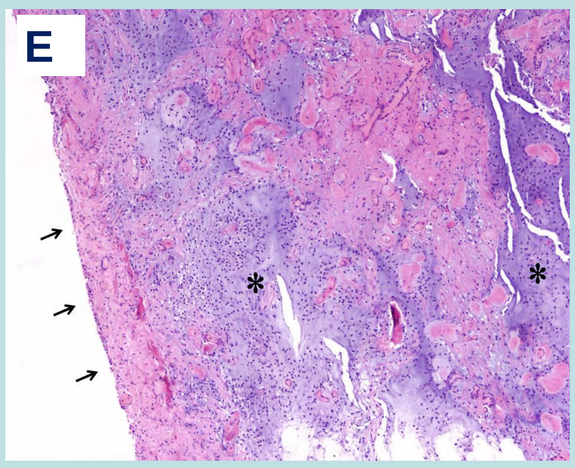
Tumour metastasis



Lumbar puncture
Cytochemical study normal, acellular cytology and CSF tumour markers (hCG beta subunit, CEA and alpha-fetoprotein) were negative.

Arachnoid biopsy
Revealed leptomeningeal tissue containing a nodule with tumour cells expressing GFAP and p16. Ki67 below 3%. Epithelial markers: negative. Diagnosis: pilocytic astrocytoma with leptomeningeal dissemination.

Fig. E, F and G



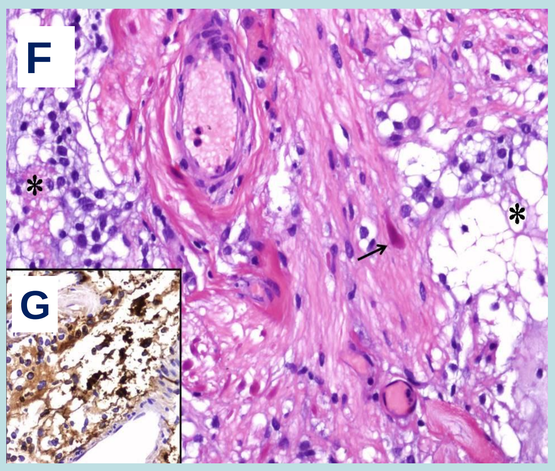


Fig. E. *Microscopic image showing the leptomeningeal surface (arrows) and a heterogeneous neoplastic proliferation alternating dense areas and loosely packed areas (*). The dense areas are formed by elongated cells on a fibrillar background. The loosely packed areas consist of round cells on a myxoid background adopting a microcystic appearance.*

Fig. F. *High magnification showing the cytological characteristics: cells with bland elongated nuclei surrounding blood vessels on a fibrillar background with presence of Rosenthal's fibres (arrow). Microcystic areas with round cells without cytological atypia (*).*

Fig. G. *Detail (left bottom): tumoral cells showing strong positivity for glial fibrillar acidic protein.*

Treatment
While awaiting the response to induction chemotherapy (SIOP-LGG 2004 protocol / Vincristine + carboplatin), treatment with LHRH analogues was not started. When the initial cycle was concluded after 2 months, a new LHRH test showed persistent HH- gonadal axis activity, and treatment with LHRH analogues was then initiated.

CONCLUSION

Hypothalamic and infundibular involvement of pilocytic astrocytoma and its association with central precocious puberty are exceptional. This entity should be considered in the differential diagnosis of infiltrative processes of the hypothalamus and pituitary infundibulum.