Symptomatic osteolipoma of the tuber cinereum

Case report

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The authors present a case of an ossified lipoma located at the tuber cinereum. Intracranial osteolipomas are rare lesions that occur in the region of the tuber cinereum. Almost all reported cases have been incidental autopsy findings. This report, however, involves a young girl who presented with an ovarian cyst and signs of precocious puberty, in whom a typical osteolipoma was surgically removed. This is the first case of hypothalamic osteolipoma presenting with endocrinological disturbances that has been published to date.

KEY WORDS • lipoma • osteolipoma • endocrine dysfunction • tuber cinereum • supraorbital approach

Intracranial lipomas are neither hamartomas nor true neoplasms; they are congenital malformations. Although they can occur anywhere in the intracranial space, a large proportion of lesions tend to be located around the midline. Interhemispheric lipomas are the most common (45%); the remainder of the lesions are found in the quadrigeminal/superior cerebellar region (25%), suprasellar/interpeduncular region (14%), cerebellopontine angle (9%), and sylvian cisterns (5%). Fifty-five percent of these lesions are associated with brain malformations of varying severity.

Osteolipomas are distinguished from other intracranial lipomas by their arrangement of central adipose and peripheral osseous tissues and by their characteristic location in the suprasellar/interpeduncular region. Ossification has been reported in many (13 [42%] of 31) suprasellar/interpeduncular lipomas. In contrast, ossification of lipomas at other intracranial sites is relatively rare. Most of the cases reported to date have been incidental autopsy findings.

We report the endocrinological, MR imaging, surgical, and pathological findings in an 8-year-old girl who underwent microsurgical removal of her benign lesion.

Case Report

History and Examination. This 8-year-old girl was referred to our pediatric neurosurgical service on March 30, 1998 for consideration of surgical management. Her medical history was unremarkable. Her first symptom was breast development. The patient had been admitted to the Department of Gynecology at Semmelweis University on March 20, 1998 for evaluation. Diagnosis of breast development was obvious at physical examination. An endocrinological evaluation performed February 12, 1998 had yielded the following results (normal ranges shown in parentheses): FSH, 1.3 U/L (4–13 U/L); LH, 0.44 U/L (1–18 U/L); progesterone, 2.8 ng/ml (0.27–2.61 ng/ml); PRL, 14.6 ng/ml (1.4–24.2 ng/ml); testosterone, 0.6 nmol/L (0.9–4.5 nmol/L); estradiol, 146 pg/ml (39–189 pg/ml); and dehydro-3-epiandrosterone, 1.7 ng/ml (normal). Ultrasonography studies of the abdomen revealed a large (4 × 5–cm) ovarian cyst on the right side. An MR image of the head revealed a lipoma-like lesion on the right side of the pituitary stalk, which extended from the tuber cinereum to the right mammillary body (Fig. 1). A differential diagnosis of a hormone-producing ovarian tumor and a pituitary lesion was made from the MR images, indicating a neuroendocrinological origin.

Operation. Surgery for removal of the lesion was chosen based on the endocrinological results and also to obtain material for a histological diagnosis. In April 1998 the patient underwent a supraorbital microcraniotomy on the right side. The skin was incised along the upper line of the eyebrow and a small (2 × 3 cm) craniotomy was made just on the frontal base. A curved dural flap was turned to the base, and after a slight retraction of the frontolateral area, the basal portion of the sylvian fissure and the optochiasmatic cistern were opened. The cerebrospinal fluid flow provided enough room to follow the structures between the internal carotid artery and the optic tract. A yellow mass was seen beneath the chiasm on the right side of the pituitary stalk. Small vessels passing over the surface of the lesion in the
The direction of the hypothalamus were cautiously separated and retracted in the anteroposterior direction. Although the lesion adhered tightly to the hypothalamus between the tuber cinereum and the right mammillary body, a good cleavage was found, and this lipomatous lesion was removed from the hypothalamus. There was no need to manipulate above the chiasm or in the midline, and therefore the lamina terminalis was not opened. The anatomical integrity of the pituitary stalk and the optic pathway were preserved.

Postoperative Course. The patient’s postoperative course was uneventful and she was discharged home on the 7th postoperative day. There was no need for hormone replacement therapy. Results of the neurological examination were unremarkable, and follow-up MR images demonstrated no residual lesion (Fig. 1).

A follow-up ultrasonography study of the abdomen obtained 2 months later demonstrated complete disappearance of the patient’s right ovarian cyst. The progress of her breast development had also stopped.

Results of follow-up endocrinological studies were normal on three separate dates. On April 22, 1998 the following hormone concentrations were recorded: FSH, 0 U/L; LH, 0 U/L; progesterone, 0.2 ng/ml; PRL, 31.3 ng/ml; estradiol, 7.3 pg/ml; and dehydro-3-epiandrosterone, 1.7 ng/ml. On November 11, 1998 the concentrations included: FSH, 2.8 U/L; LH, 0 U/L; progesterone, 0.35 ng/ml; PRL, 43 ng/ml; testosterone, 0.5 nmol/L; sex hormone–binding globulin, 94 nmol/ml; estradiol, 0 pg/ml; and thyroid-stimulating hormone, 2 mU/L (normal 0.49–4.67 mU/L). On September 13, 1999 the concentrations were: FSH, 4.55 U/L; LH, 0.07 U/L; PRL, 25.3 ng/ml; and thyroid-stimulating hormone, 2.546 mU/L. The transitory elevation of the PRL level was probably related to the surgical procedure. Two years later the girl is doing well, with appropriate somatic and mental maturation; menarche has not yet occurred.

Pathological Findings. The pathological findings proved that the lesion was a centrally located mature adipose tissue surrounded by fibrous tissue and mature bone (arrow). The lesion is characteristic of what has been called an osteolipoma of the tuber cinereum (Fig. 2).

Discussion

Tumor Origin

Intracranial lipoma formation resulting from abnormal persistence and maldifferentiation of the meninx is a widely accepted concept. The relative frequencies of the locations of the lipomas correspond to the temporal sequence of dissolution of the meninx primitiva, the mesenchymal anlage of the meninges. This finding supports the concept of the embryological development of intracranial lipomas and explains the high frequency of callosal and other brain hypoplasias. Traditionally, osteolipomas of the tuber cinereum have been classified as meningeal lipomas that are located at that site. These lesions exhibit a remarkable con-
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Slight variation in their size and in their location between the mammillary bodies and the pituitary stalk, and approximately half of them contain a well-organized bone sphere with a central marrow space. In these respects the lesions differ from meningeal lipomas in general; reasons have been proposed for considering that osteolipomas are displaced elements of the mesenchyme encompassing walls of the craniopharyngeal duct.

Tumor Symptoms

A review of the literature shows that these lesions are mostly asymptomatic. When symptoms occur, they are frequently the result of coexisting general clinical conditions. Advances in neuroimaging methods have vastly improved the likelihood of the tumor being discovered while the patient is alive. At present, however, this case is the first revealed by endocrinological signs. A possible explanation for the pathological hormone levels could be compression of the pituitary stalk and the tuber cinereum, which results in lower levels of inhibiting factor. Of course we cannot exclude the possibility of the unrelated coexistence of a hypothalamic pathological condition and its attendant endocrinological disturbances, but the complete postoperative normalization of the hormone levels supports the first hypothesis. The elevated postoperative PRL level could be explained by the minimal and transient vascular damage caused by the microdissection at the level of the tuber cinereum.

Indications for Surgery

Surgical removal of the lesion is not considered necessary in asymptomatic patients. On the other hand, when clinical signs, as in our case, seem to be related to the compression caused by the lesion, surgery is indicated. In these instances, the minimally invasive surgical approach is mandatory. The supraorbital craniotomy allows enough room for safe microsurgical dissection around the pituitary stalk. In patients of this age the small size of the frontal sinus helps to avoid the possible complication of cerebrospinal fluid leakage. With careful skin closure there is no scarring.

We recommend this approach for the exploration of the suprasellar region in young children as well as in adults.

References


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