Malignant teratoma arising within a lipomeningocele

Case report

J. PARKER MICKLE, M.D., AND JAMES E. McLENNAN, M.D.

Department of Neurosurgery, Children's Hospital Medical Center, Boston, Massachusetts

The authors report the case of an 11-year-old, neurologically normal girl, followed since birth for lipomeningocele, who developed a malignant teratoma within the lumbosacral lesion and presented with metastases to the lungs and groin. This rare occurrence is discussed from an embryological and management viewpoint.

KEY WORDS • spina bifida • malignant teratoma • embryology

S oft-tissue lumbosacral masses with underlying spina bifida are not common findings in newborn infants. These lesions are often treated conservatively so as not to risk significant neurological deficit from attempted surgical repair; this is particularly important if the child has normal neurological function. Malignant tumors associated with dysraphic malformations are quite rare. This report illustrates such a case and emphasizes the importance of recognizing morphological changes in these presumably stable lesions.

Case Report

This 11-year-old girl was born with a spongy sacral mass measuring $6 \times 5$ cm, which was thought to be a lipoma. At 2 months, neurological examination was normal. X-ray films of the lumbosacral spine revealed spina bifida of the L-4, L-5, and S-1 vertebrae, with an overlying homogeneous mass.

She was followed with an annual examination, and remained entirely normal except for mild lumbar scoliosis and a 0.5-cm leg length discrepancy. Five months before a scheduled visit to the neurosurgical clinic, the patient and her parents noted a more solid mass within the lipoma. A right groin mass also appeared and increased slowly in size over the ensuing months. No symptoms were present except for localized discomfort with pressure on the sacral mass. Retrospectively, the patient's mother had always been aware of a small solid component to this lesion.

Examination. The patient had a nonpulsatile, superficial, nontender mass in the right groin which was mobile and firm, and measured $1.5 \times 3$ cm. There was also a soft, $10 \times 12$-cm, sacral lipoma containing a firm,
movable 6 × 8-cm mass over the right lumbosacral junction. Palpation of this mass produced no symptoms. Orthopedic examination showed a mild scoliosis with a lumbar convexity to the right and a 0.5-cm shortening of the left leg. Neurological examination was entirely normal as was bladder function.

Femoral arteriography ruled out a vascular lesion of the groin, and the lump was excised. Histologically, this proved to be a mass of primitive cells with mesenchymal differentiation described as an "embryoma." Gas myelography showed the feasibility of removing the sacral mass; this study demonstrated a low-lying conus medullaris and a small focal meningocele at L5–S1 protruding only as far posteriorly as the dorsal spines of these vertebrae (Fig. 1).

Metabolic and metastatic workup included normal roentgenographic bone survey, liver and spleen 99Tc scans, lymphangiography, intravenous pyelography, vanillylmandelic acid determination and serum enzyme levels. Chest tomograms demonstrated several small peripheral nodules thought to be metastatic tumor.

Operation. The firm sacral mass was approached through a transverse incision across the lipoma. A tough vascular tumor was encountered, attached to the presacral fascia and external to the meningocele; it was totally excised. Histologically, this tumor resembled the groin lesion but also showed characteristics of a teratocarcinoma; the final pathological diagnosis was Wilms' tumor (Fig. 2). Postoperative neurological examination remained normal.

Postoperative Course. A combined course of actinomycin-D, cytoxan, vincristine and adriamycin was initiated on the seventh postoperative day and the patient was discharged on the tenth day. Except for a mild reaction to the chemotherapy, which has thus far resulted in disappearance of the lung metastases, the patient remains well.

Discussion

Malignant degeneration within a congenital malformation of the distal neural plate is a rare phenomenon. Thorp and Love each reported a case of spina bifida with delayed malignant transformation. Thorp's patient was a 26-year-old man with a myelomeningocele and chronically draining dermal sinus eventuating in an anaplastic car-
Malignant teratoma arising within a lipomeningocele

cinoma. The other patient was a 30-year-old man with a spina bifida and a lumbosacral lipomeningocele. The mass became firm and tender and a right inguinal mass appeared; excisional biopsy of the sacral lesion showed a malignant teratoma with extensive adenocarcinoma. Both patients died only months after diagnosis.

The present case closely resembles that reported by Love. Both previous reports, however, emphasize chronic irritation as a major etiology of malignant degeneration within the dysraphic lesion. This factor is absent in our case.

A most intriguing study was presented by Gruenwald. A 20-mm human embryo with myeloschisis was found to have regions of dystotic tissue (primitive mesenchyme, ectopic cartilage anlage, nephric glomeruli) within the caudal portion of the embryonic neural malformation. Perhaps the delayed appearance of malignancy in our patient represents an extreme of the spectrum of malformation resulting from the multipotent caudal neural primordium. It is possible that incipient adolescent endocrinological changes activated a dormant teratomatous deposit.

It is stressed that patients with presumably benign lipomeningoceles should be followed expectantly for the rare but disastrous occurrence of delayed malignancy.

References


This work was supported in part by grant 5T01-NS05519, from the National Institutes of Health.

Address reprint requests to: J. Parker Mickle, M.D., Department of Neurosurgery, Children's Hospital Medical Center, Boston, Massachusetts 02115.