Primary cutaneous extravertebral meningioma

Case report

MENASHE ZAAROOR, M.D., BERNARDO BOROVICH, M.D., LUCYNA BASSAN, M.D., YAFFA DORON, M.D., AND JAN GRUSZKIEWICZ, M.D.
Departments of Neurosurgery and Pathology, Rambam Medical Center, Technion Faculty of Medicine, Haifa, Israel

A case of cutaneous extravertebral meningioma is presented. It was diagnosed in infancy as a lumbar meningocele. Operation was initially refused but was subsequently demanded for cosmetic reasons. The findings were a very thick corrugated skin and a cutaneous meningioma connected by a fibrous tract to the dura mater. The presence of a fibrous stalk linking the tumor to the dura mater might have been the pathogenetic connection between the meningocele and cutaneous meningioma.

KEY WORDS • meningocele • cutaneous tumor • meningioma

MENINGIOMA is one of the commonest primary tumors of the central nervous system. Only very occasionally have extracranial or extravertebral meningiomas been reported. They can be secondary to extension of an intracranial or intraspinal tumor or they can be primary. The most common sites of these primary extracranial or extraspinal meningiomas are the orbit, the vicinity of the ear, and the skin. Meningiomas of the skin are the rarest and have been reported in the scalp, the forehead, and the paravertebral region, where its occurrence is exceptional. Lopez, et al., in their extensive review, found only two cases of primary cutaneous extraspinal meningiomas, thus justifying our report of an additional example.

Case Report

This 13-year-old girl was admitted to the Rambam Medical Center on November 29, 1981, because of a lump in the right paravertebral lower lumbar region just off the midline. It was present at birth, but an operation was refused at that time, and the mass had not changed with time. Surgery was now requested for cosmetic reasons.

The swelling (Fig. 1) measured 5 cm in diameter, had a narrow neck, and was firm and rubbery. The skin was hard and corrugated. Transillumination test was negative, and neurological examination was normal. Plain radiography of the spine showed a bifid L-4 vertebra. At surgery, the swelling was resected. The hard irregular skin accounted for half the thickness of the mass; the rest was subcutaneous fat. A meningocele was not found; instead, a fibrous tract, 0.5 cm in diameter, made its way through the interlaminar space and linked the bulky skin to the intraspinal dura mater. Postoperative recovery was uneventful.

Microscopic examination revealed a fragment of skin with a hyperkeratotic epidermis. The upper dermis

FIG. 1. Paravertebral mass in the lower lumbar region just to the right of the midline.
showed mild focal lymphocytic infiltration. In the deeper layer of the dermis and subcutaneous tissue, nests and sheets of polygonal cells were seen with large spherical nuclei, which had a fine chromatin pattern. These areas were separated by wide bands of collagen. In some regions, typical meningeal whorl formations, calcified psammoma bodies, and so-called “collagen bodies” were present (Fig. 2).

Discussion

Primary cutaneous meningioma is a very rare tumor. As defined by Lopez, et al., it occurs in children, is asymptomatic, and is present at birth. The preferential sites are the scalp, close to the midline along the cranial sagittal suture, and the skin, at the glabella and paravertebral region. The latter location is exceptional. In their extensive review, Lopez, et al., found only two cases.

These lesions are thought to be the result of a developmental defect. Meningeal cells get sequestered or trapped in the skin at the time of closure of the neural crest. The same defective closure of the neural crest underlies the formation of meningoceles and primary cutaneous meningiomas. Both can appear in the same lesion. The cutaneous tumor can be an isolated finding and have no direct relation to the central nervous system or its coverings. The present case may represent an intermediate stage in the chain of events linking meningocele and cutaneous meningioma: a fibrous stalk instead of a meningocele connected the cutaneous swelling to the intraspinal dura mater through an L-4 bifid spine. This fibrous tract would be what Lopez, et al., interpreted as a spontaneously obliterated meningocele.

References


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Address reprint requests to: Menashe Zaaroor, M.D., Department of Neurosurgery, Rambam Medical Center, Haifa, Israel.