Unusual case of extradural choroid plexus papilloma of the sacral canal

Case report

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An unusual case of a sacral, extradural choroid plexus papilloma involving the S1–3 level is described. This 50-year-old woman presented with a 4-month history of pain involving her right buttock, perineum, and leg. Contrast-enhanced magnetic resonance (MR) imaging of the spine revealed a well-defined, mildly enhancing sacral canal mass at the S1–3 level; its appearance was consistent with that of a benign tumor. Intraoperatively, the lesion was found to be extradural in location and was entwined among nerve roots in the sacral canal. Microscopic examination of the gross totally resected tumor revealed typical features of a choroid plexus papilloma. Despite performing a thorough neuroimaging workup (craniospinal contrast-enhanced MR imaging) for an intracranial or spinal primary mass, none was found. The choroid plexus appeared entirely normal; however, both a cavum septum pellucidum and a cavum vergae were noted. Extraneural choroid plexus papilloma, specifically intrasacral, extradural choroid plexus papilloma has not been previously reported. The present example is thought to have arisen either from ectopic choroid plexus tissue or perhaps by metaplasia from ependymal rests.

KEY WORDS • choroid plexus papilloma • ectopic neoplasm • maldevelopment • embryology

Abbreviations used in this paper: CPA = cerebellopontine angle; CSF = cerebrospinal fluid; MR = magnetic resonance.

Case Report

Examination. This 50-year-old woman with no history of tumor presented with a 4-month history of increasing right buttock, perineum, and posterior leg pain. Physical examination disclosed right perineural and perianal numbness. Sensory abnormalities were also noted in the S-3 and S-4 dermatomes of the right leg. No motor deficits were noted. Reflexes were normal and no long track signs or cranial nerve dysfunction was observed. A rectal examination revealed poor sphincter contraction on the right.

Neuroimaging. A contrast-enhanced spinal MR image demonstrated a solitary, heterogeneously enhancing mass at the S1–3 level (Fig. 1). Its circumscription and general appearance were consistent with a benign tumor, thus a presumptive diagnosis of schwannoma or meningioma was made.

Operation. A sacral laminectomy was performed, which revealed a soft gray tumor lying within the sacral canal at the S2–3 levels, well below the level of the “arachnoid bubble” and outside the dural sac. It was intimately entwined with and compressed the dura-enveloped sacral nerve roots. Despite accomplishing an extensive resec-
tion, minute gelatinous-appearing foci of presumed tumor were evident among the nerve roots that were viewed under the dissecting microscope.

Postoperative Course. Cranial and spinal contrast-enhanced MR imaging was performed to exclude the presence of a primary tumor at a higher site. No tumor was identified at 8-month follow-up examination, but another contrast-enhanced MR image was obtained that revealed foci of enhancement in the tumor bed. Multiple biopsy samples revealed only scar tissue; no residual tumor was found. The initial removal was therefore considered to be a total resection. At 9-month follow-up visit, the patient has made a near-complete recovery. Bowel and bladder function were restored; only patchy numbness remained in the S3–5 distribution.

Pathological Findings. Microscopy revealed a well-differentiated papillary epithelial neoplasm, which consisted of a single layer of cuboidal to columnar cells resting on a papillary fibrovascular stroma containing occasional calcifications (Fig. 2). No goblet cells were seen within the epithelium and no adipose or other mesenchymal tissues were identified. Cytological atypia and mitotic activity were lacking. Nonneoplastic choroid plexus epithelium was not found. Using the streptavidin-biotin-peroxidase complex method, the lesion was found to be immunoreactive for S-100 protein (polyclonal, dilution 1:800; Dako Corp., Carpinteria, CA), cytokeratin (clone CAM 5.2, dilution 1:50; Becton Dickinson, San Jose, CA), vimentin (clone 3B4, dilution 1:500; Dako Corp.), and synaptophysin (clone SY38, dilution 1:750; ICN, Costa Mesa, CA). The glial fibrillary acidic protein preparation showed patchy but distinct staining of tumor cells. Epithelial membrane antigen (clone E29, dilution 1:20; Dako Corp.) as well as estrogen receptor (clone 6F11, predilute; Ventana Medical Systems, Tucson, AZ) and progesterone receptor (clone 1A6, prediluted; Ventana Medical Systems) preparations were all negative. The Ki-67 labeling index (clone MIB-1, diluted 1:400; Immunotech-Coulter, Hialeah, FL) was 5%. Based on these morphological and immunohistochemical findings, a diagnosis of choroid plexus papilloma was confirmed.

Discussion

Choroid plexus tumors arise primarily within the ventricular system, where the choroid plexus is normally found. Most occur in either the lateral or the fourth ventricles. Third ventricular examples are much less common. Only occasional papillomas are extraventricular in origin. Tumors occurring in the CPA represent either direct extension of fourth ventricular choroid plexus papilloma via the recess and foramen of Luschka or arise from the tuft of choroid plexus normally protruding from that foramen. Only an occasional choroid plexus papilloma located in the CPA is independent of the normal choroid plexus at that site. Kimura, et al., reported a primary choroid plexus papilloma in the suprasellar region, a tumor having no connection with the ventricular system. Such lesions are thought to arise from embryonic remnants of choroid plexus tissue. In patients with Chiari Type II malformations, ectopic fibrovascular tissue-containing choroid plexus may be seen between the vermis and medulla. Ectopic, apparently secreting choroid plexus tissue has also been described in occasional cystic lesions without ventricular communication. These include arachnoid cyst of the CPA, intracerebral supratentorial cyst, and oropharyngeal cyst.

Although choroid plexus papillomas are histologically benign, they have been reported to undergo cerebrospinal seeding to distant sites, both intracranial and spinal. Reported cases of spinal spread clearly indicate that dissemination occurs via CSF and involves the subarachnoid space. This seeding process appears to be independent of malignant transformation, because the deposits retain their benign histological and ultrastructural appearance. Wolfson, et al., reported an example studied by electron microscopy in which the “metastatic deposit” very closely resembled normal choroid plexus.

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Fig. 1. Sagittal (left) and axial (right) contrast-enhanced MR images revealing the solitary, circumscribed tumor at the S1–3 level to be heterogeneously enhancing. Note its intimate association with nerve roots in the sacral canal.

Fig. 2. Photomicrographs. Left: The tumor showed classic features of choroid plexus papilloma, including crowding of papillae with a delicate fibrovascular stroma. Right: The papillae were lined by a single layer of largely columnar epithelial cells lacking atypia or mitotic activity. No other tissue elements were noted.

Despite the unusual site of occurrence of the present lesion, its histological and immunocytochemical features entirely fulfill the diagnostic criteria for choroid plexus papilloma. Because the morphological features of choroid plexus papilloma may mimic those of normal choroid plexus, it could be argued that the lesion represents a teratoma with predominance of a single tissue type (monodermal teratoma). Although choroid plexus is a relatively common component of central teratomas, the specimen possessed no other tissue elements to suggest that diagnosis. In any event, sacral teratomas are very rare; the same is true of spinal intradural examples. Metastatic papillary carcinoma may also be considered in the differential diagnosis. The well-differentiated morphology of our lesion, its benign cytology, lack of mitotic activity, and low-level MIB-1 labeling index all argue against that diagnosis, as does S-100 protein, synaptophysin, glial fibrillary acidic protein, and vimentin immunoreactivity coupled with lack of epithelial membrane antigen and carcinoembryonic antigen staining. Strong staining for synaptophysin has only recently been shown to distinguish normal choroid plexus and its tumors from metastatic carcinoma. Finally, no intercranial or other intraspinal primary tumor was found on postoperative MR images.

Both cavum septi pellucidi and cavum vergae are midline developmental abnormalities of the brain. Although they are frequent findings in premature and full-term infants, a persistent cavum septum pellucidum is encountered in only 2 to 4% of normal adults. The presence of these developmental anomalies in association with an ectopic choroid plexus papilloma raises the question of whether the tumor had its origin in a malformation, such as ectopic choroid plexus. As previously noted, the specimen in our case was devoid of normal choroid plexus.

Although the tumor in our patient arose within the sacral canal rather than intradurally, the complex embryogenesis of this region must be taken into consideration as a possible basis for its tumorigenesis. During the process of spinal cord embryogenesis, the cervical, thoracic, and lumbar segments develop from neural tube by the process of primary neurulation. On the other hand, saccroccygeal segments develop from the caudal end by secondary neurulation. The latter process is followed by formation of the terminal filum, ventriculus terminalis (fifth ventricle), and the conus medullaris by so-called canalization with retrogressive differentiation. These structures all develop from a cluster of undifferentiated pluripotential cells called the caudal cell mass. Authors of one report on five intradural lipomas or lipomyelomeningoceles suggested that congenital lipomas of the lower spinal canal are formed by persistence and differentiation of these pluripotential cells. The authors of the same report also described various ectopic neuroectodermal and mesodermal tissues within the lipomatous stalk of these lesions. These included neuroglia, ependyma, and smooth as well as striated muscle. The finding of such ectopic tissues may signify failure of retrogressive differentiation. Perhaps the occurrence of choroid plexus epithelium, the usual precursor of choroid plexus papilloma in this general region, could be explained by the same mechanism. The possibility that choroid plexus epithelium might have arisen by metaplasia from ependymal rests, the same that give rise to occasional ectopic extraneural myxopapillary ependymomas in the lumbosacral region, cannot be excluded. Support for this notion can be found in the histogenetic relationship of these tissues, the frequent occurrence of ependymal differentiation in choroid plexus papilloma, and the rare occurrence of choroid plexus differentiation in ependymomas.

In summary, we have reported a unique example of choroid plexus papilloma in the sacral canal. Careful gross-total resection without adjuvant therapy may be sufficient treatment for such lesions. Given the classic histology of our tumor, the absence of a neoplastic process at other sites in or outside the central nervous system, and the association with minor central nervous system malformations, we believe it was primary at this site and either arose in an ectopia or by way of metaplasia from ependymal rests.

References

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