Schwannoma with monoclonal plasma cell infiltration

Case report

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The authors report a very rare case of a vestibular schwannoma with an infiltrate of monoclonal plasma cells. A 45-year-old woman underwent routine excision of a presumed vestibular schwannoma. Histological analysis revealed the presence of a distinct lambda light chain restricted plasma cell population within the schwannoma. The light chain restriction and polymerase chain reaction–demonstrated monoclonality of the plasma cell population suggested the co-occurrence of a plasma cell neoplasm within a schwannoma. A search for systemic disease of plasma cell origin was unremarkable. A search of the literature suggests that this is the first report of such an occurrence.

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KEY WORDS • vestibular schwannoma • plasmacytoma • collision tumor • plasma cell

Examination. This 45-year-old woman with moderate to severe learning difficulties presented with ataxia that had increased over a number of years. She lived in a supported care home, and until the onset of symptoms she had been independently mobile. She initially presented to the neurology services with headaches and left-sided trigeminal neuralgia. Her dizziness had become so severe that she was now wheelchair bound. Examination also revealed reduced sensation over the left side of her face. A CT scan showed a large mass in the posterior fossa, and an MR image confirmed the presence of an extensive mass in the left cerebellopontine angle compressing the brainstem and consistent with a vestibular schwannoma (Fig. 1).

Operation and Postoperative Course. The patient underwent subtotal excision in which the mass was approached via a retromastoid approach. The tumor was noted to be highly vascular with a rubbery consistency. She made an uneventful recovery from surgery and was able to return to her care home.

Pathological Findings and Further Management. Histological examination demonstrated that most of the lesion was composed of interlacing fascicles of spindle-shaped cells with Verocay body formation; the features were diagnostic of a benign WHO Grade I schwannoma (Fig. 2). There were also smaller areas containing poorly delineated nodules of hypercellularity containing an infiltrate of atypical plasma cells showing lambda light chain restriction (Figs. 3 and 4). Both CD56 and Cyclin D1 were negative. The MIB-1 labeling index was 6% consistent with a plasma cell neoplasm. Polymerase chain reaction studies demonstrated monoclonality of the variable region of the immunoglobulin-heavy chain. The features were suggestive of a neoplastic plasma cell population within a schwannoma, and therefore the diagnosis of plasmacytoma and schwannoma was made.

A limited search for systemic plasma cell disease was conducted because of the patient’s comorbidities and quality of life, and it was therefore decided that a bone marrow biopsy was not appropriate in this case. However, screening CT scanning did not demonstrate any evidence of disseminated plasmacytoma, and both Bence Jones proteins and discrete bands were absent on plasma
electrophoresis. Gamma globulin levels were normal. Ordinarily the search for systemic plasma cell disease would have continued for at least 1 year. Localized intracranial plasmacytoma would have been managed with radiotherapy with potentially very good results.

Discussion

Cerebellopontine angle tumors account for ~10% of intracranial tumors, and vestibular schwannomas account for the vast majority (up to 80%) of these. The presence of 2 or more tumors of different origins is a well-recognized phenomenon, occurring in 8% in some autopsy studies. However, the presence of 2 tumors in the same anatomical location is much rarer. It is thought that in such cases 1 tumor has metastasized into the other. Tumor-to-tumor metastasis or “collision” tumors are characterized by the presence of 2 parts, 1 at least partially enveloping the other, and of distinct histological character and origin.

While inflammation is a frequent feature of vestibular schwannomas in histological specimens, in the present case the large number of monoclonal plasma cells within the tumor was strong evidence that this lesion represented a separate tumor of plasma cell origin.

Plasma cell tumors may either be isolated monoclonal proliferations of plasma cells (solitary plasmacytoma) or may be a disseminated systemic disease (multiple myeloma). In this patient no evidence of systemic disease has been found, but in the absence of bone marrow biopsy, it cannot be considered to have been excluded.

There have been only a few reported cases of cerebellopontine angle collision tumors in the literature, and none has involved plasmacytomas. The hosts are either meningioma or vestibular schwannoma, and typical metastases include breast or lung carcinoma or melanoma. One suggestion is that these slow-growing, benign tumors, which are frequently highly vascular, form excellent non-competitive environments for subsequent metastases.

Intracranial plasmacytoma is an uncommon condition, making the present case even more unusual. Solitary plasmacytoma of bone has been described at the skull base and may affect the petrous bone, and extramedullary plasmacytomas may affect the nasopharynx. While the progression of isolated plasmacytoma to myeloma is more common in solitary plasmacytoma of bone than extramedullary plasmacytoma, progression of intracranial plasmacytoma is more common if it affects the skull base, and Schwartz et al. have recommended continuing a search for systemic disease for up to 1 year after initial diagnosis.

This case is an example of a very unusual collision...
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A tumor consisting of intracranial plasmacytoma and a vestibular schwannoma of the cerebellopontine angle. Such occurrence has not previously been reported in the literature.

**Disclaimer**

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