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Case report

Schwannoma of right cerebellopontine angle. A cytologic diagnosis

Alexandra Kalogeraki[1], Dimitrios Tamiolakis[1], Iris Zoi[1], John Segredakis[1], Maria Kafousi[1], Antonios Vakis[2]

[1] Department of Pathology-Cytopathology, [2] Department of Neurosurgery, Medical School, University of Crete, University Hospital, Heraklion, Crete, Greece;

Summary. Schwannomas affect mainly head and neck peripheral nerves, are benign tumors and derive from Schwann cells. Schwannoma of right cerebellopontine angle is extremely rare to diagnose by cytology. We report one such rare case presenting the cytological features in material obtained during the resection of the tumor. *Case report:* A 47-year-old female was diagnosed by MRI with a tumor of right cerebellopontine angle.. Cytologic material from the tumor was obtained intraoperatively and diagnosed cytologically as a neurilemoma. *Conclusion:* This case is presented here to focus the ability of cytology in diagnosis of schwannoma in intraoperative material of the tumor, using immunohistochemistry and confirmed by histology- immunohistochemistry. (www.actabiomedica.it)

Key words: Schwannoma, cytology, histopathology, immunocytopathology

Introduction

Neurilemomas or Schwannomas are the most common benign encapsulated neoplasms arising from peripheral nerve tissue. Usually they are encapsulated perineural tumors of neuroectodermal derivation that originate from the Schwann cells of the neural sheath of motor and sensory peripheral nerves The etiology is still unknown (1,2).

Schwannomas occur in patients with neurofibromatosis type 2 (NF2) and schwannomatosis. Most NF2 patient tumors have biallelic inactivating mutations in the NF2 tumor suppressor gene. Schwannomatosis patients do not harbor the NF2 germ line mutations, and the molecular basis of their disease remains unknown, however schwannomas from schwannomatosis patients also have biallelic NF2 mutations.

The NF2 protein, merlin or schwannomin, belongs to the ERM (ezrin-moesin-radixin) family of membrane-cytoskeleton linking proteins. There is a loss of NF2 in schwannomas. NF2 inactivation causes tumor formation through cellular changes and subsequent cytoskeletal abnormalities (13).

Neurilemomas occur in all age groups, but most frequently between 20 and 50 years. Many patients have minor symptoms from the tumor and pain after surgery can be more pronounced than it was before operation. When an excision of the lesion is considered necessary a correct diagnosis helps the surgeon to plan surgery so as to avoid neurological sequelae (3).

This case has been reported for its rare and unusual site for cytodiagnosis of a Schwannoma confirmed by histology.

Case report

A 47-year-old female patient was presented at Univ. Hospital of Heraklion Crete and diagnosed by MRI with a solid- cystic tumor of cerebellum. She

suffered from cranial pain and there was no history of trauma. Past, personal and family history was non-contributory.

The hematological and biochemical parameters were with in normal limits. The MRI signal in the center of the mass was hyperintense on T2-weighted and isointense on T1-weighted images.

Material and Methods

Cytology: The cytologic material obtained intraoperative (during the resection of tumor) was smeared on glass slides. The air dried smears were used for Giemsa stain and immunochemistry while the alcohol (80%) fixed for routine Papanicolaou stain (Fig. 1).

Immunocytology: In air dried smears immunocytochemistry was performed using the markers S-100 protein (Fig. 2) and Vimentin

Histology: In histological specimens of the tumor fixed in 10% formalin, the H-E stain was performed (Fig. 3).

Immunohistochemistry: The markers S-100 protein, Vimentin, EMA, NF, GFAP, Calretinin, PGM-1 and the proliferation index MIB-1 were used.

Results

Cytology: showed cellular smears of spindle shaped tumor cells and Schwann cell processes. Isolated cells were elongated, round to spindle shaped with elongated, slender vesicular nuclei with ill-defined cytoplasm. In the background many lymphocytes and histiocytes were found.

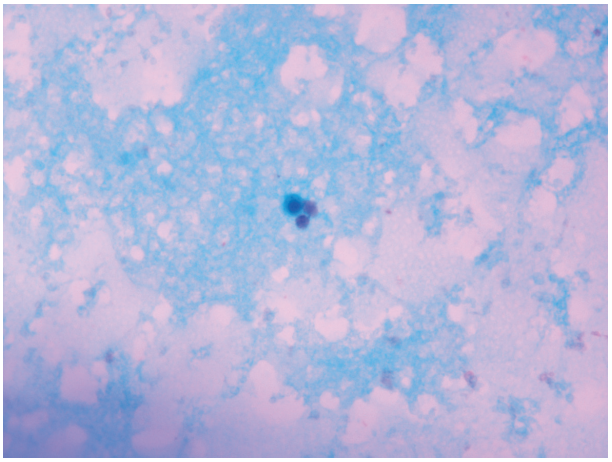

Figure 1. Schwannoma, Intraoperative smear. Isolated neoplastic cell. Papanicolaou stain X400

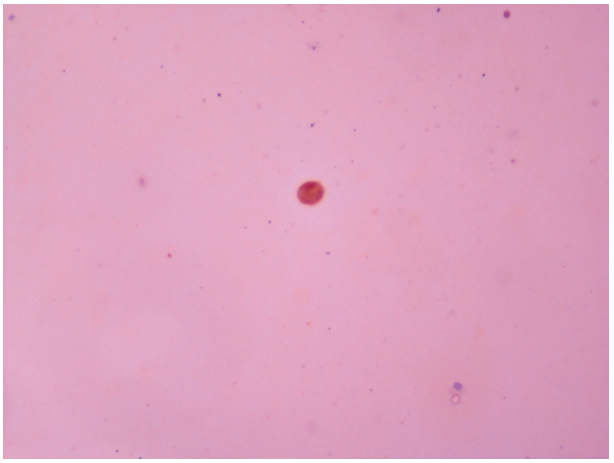

Figure 2. Schwannoma, Intraoperative smear. Isolated neoplastic cell. S-100 immunostain X400

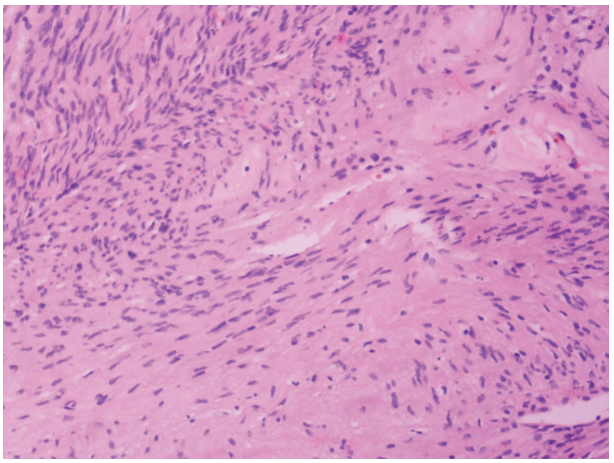

Figure 3. Schwannoma. Tumor section. Hematoxylene-Eosin(H@E) stain X 400

Histology: Abundant material with many spindle-shaped neoplastic cells with mild pleomorphism, rare nuclear atypia and nuclear inclusions. At places Verocay bodies were found and in the background many histiocytes were observed. Mitoses or necrosis were not found.

Immunocytochemistry: The majority of neoplastic cells were found to be cytoplasmic positive for S-100 protein (Fig. 4).

Immunohistochemistry: The tumor cells expressed cytoplasmic S-100 protein, Vimentin and EMA mark-

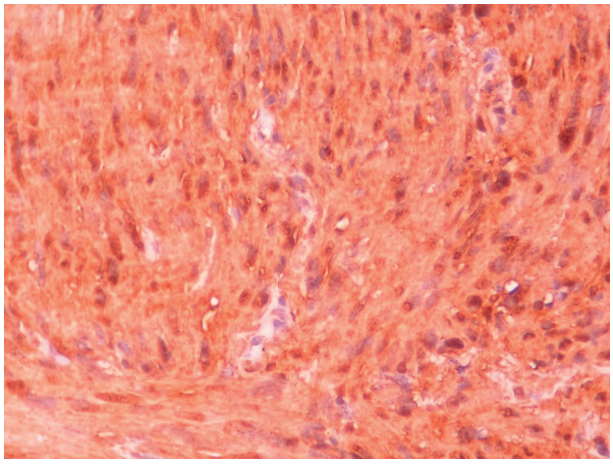

Figure 4. Schwannoma. Tumor section. S-100 immunostain X 400

ers. The neoplastic cells were found to be negative for NF, GFAP and PGM-1 and proliferating index MIB-1 was found to be positive in 1% of tumor cells.

Discussion

About 25% of the Schwannomas occur in the head and neck region (3,5), usually involving cranial nerves and sympathetic chain. The schwannoma is the most common tumor of the peripheral nerve occurring anywhere in the body (4,5), often presents as a solitary painless, and slow- growing mass of variable size.

Cystic schwannoma is thought to grow more rapidly than non- cystic (4,8,10).

Smears from schwannomas with cystic degeneration obtained preoperative by FNAB show scattered round- to- oval cells, accompanied by occasional histiocytes (3-6,8,10).

In this report the cytologic material was obtained intraoperative and was adequate characteristically composed of spindle- shaped cells, vesicular nuclei, scanty cytoplasm without mitoses and necrosis with the presence of many lymphocytes and histiocytes because of the cystic degeneration of the tumor.

Histologically, a typical schwannoma is composed of two areas (11): The Antoni A area characterized by closely packed spindle cells with occasional nuclear palisading and Verocay bodies as observed in our case. The Antoni B area is occupied by loosely arranged tumor cells which are separated by abundant myxoid stroma. An Antoni A area (cohesive cellular clusters) and an Antoni B area (loosely cohesive or poorly cellular sheets) are occasionally found in cytology (2,3).

Immunohistochemically, schwannomas are usually positive for S-100 protein,Leu-7 (CD57),and glial fibrillary acidic protein (GFAP) (7,9-12).

The immunophenotype of schwannomas is highly distinctive: S-100, Collagen IV and lamin, are expressed expecially in Antoni A areas, GFAP may be seen in a significant number of schwannomas, Neurofilament protein NFP staining is limited.

Recent markers include podoplanin, calretinin and SOX10 (14, 15).

In our case the majority of neoplastic ells were found to be cytoplasmic positive for S-100 protein but negative for Vimentin by immunohistochemistry.

In our case the schwannoma occurs in right cerebellopontine angle and in this region is a very rare entity for cytologic diagnosis of schwannoma because of FNAC aspirates have often inadequate material (2,3,7,8).

By immunohistochemistry the tumor cells expressed cytoplasmic S-100 protein, Vimentin and EMA markers but were found to be negative for NS, GFAP and PGM-1 markers.

In conclusion, hence FNAC is not helpful in achieving preoperative diagnosis in case of schwannoma, the intraoperative cytology provides a convenient and safe diagnosis confirmed by histology.

Writen informed consent was obtained by the patient to submit this case report to the journal.

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Correspondence:
Alexandra Kalogeraki, MD, PhD
University of Crete, Medical Faculty, PO Box 1393
Heraklion 71110, Crete Greece
Tel. + 30 2810 394692
Fax +30 2810 394694
E-mail: kalogerakimed@yahoo.gr