Ancient schwannoma of the neck mimicking soft tissue sarcoma

Sir,
We wish to describe a case of a 46-year-old women presenting with ancient schwannoma of the neck. Only few cases of ancient schwannoma have been reported in the neck region. In our patient, the tumor showed degenerative changes and nuclear atypia, thus mimicking a mesenchymal neoplasm.

A 46-year-old female with no symptoms or family history of von Recklinghausen disease was admitted to the department of surgery with a 6-month history of a swelling in the neck region. The swelling was non-tender, was not mobile, and the overlying skin was normal. We made a preoperative diagnosis of myolipoma Excision biopsy was done under local anesthesia and the specimen was sent for histopathological examination.

An ovoid mass of 6 × 5 cm was sent to the pathology department for gross examination. The tumor was firm, well circumscribed, and encapsulated. The cut surface showed cystic degeneration and hemorrhage. The tissue was routinely processed and sections were cut at 5 mm thickness and stained with Hematoxylin and Eosin (H and E).

The H and E stained sections of the tumor showed Antoni-A and Antoni-B areas surrounded by a fibrous capsule. The Antoni-A regions were cellular and showed Verocay bodies(eosinophilic cell bodies nearly encircled by rows of nuclei). The Antoni-B areas were hypocellular. Cells were loosely arranged in a myxoid matrix. Infiltrationby lymphocytes, mast cells, and pigment-laden macrophages,was seen in Antoni B areas. In addition cystic changes and thick wall blood vessels were unusually prominent. The Schwann cell nuclei were large, hyperchromatic, and multilobated, and exhibited marked nuclear atypia. The cytoplasm showed vacuolar changes. These findings were compatible with a diagnosis of ancient schwannoma (degenerated neurilemmoma).

Photomicrograph shows degenerative nuclear atypia (blue arrow), cystic changes (green arrow) and thick walled congested blood vessels (black arrow) (H&E, ×400).

Schwannomas are encapsulated benign tumors arising from nerve sheath cells; the ancient schwannoma is one of the five variants.[1] Ancient schwannomas are neurilemmomas that display pronounced degenerative changes. Ancient change in a schwannoma is a histological variant that is typically found in long-standing tumors, and is thought to result from degenerative changes.[2] Degenerative changes include cyst formation, calcification, hemorrhage, and hyalinization. One of the most misleading aspects of this tumor is the degree of nuclear atypia. These tumors behave as ordinary neurilemomas and therefore the nuclear atypia can be regarded as a purely degenerative change.[3]

Ancient schwannoma of the neck region is a rare benign neoplasm derived from neural crest cells and is usually solitary., only a few ancient schwannomas have been reported in different locations in the neck region.[1] A significant percentage of ancient schwannomas are located in deep locations such as the retroperitoneum.[2] Ancient schwannomas of the neck are frequently misdiagnosed, and preoperative investigations are therefore often fruitless.[4] The histopathological features such as degeneratve changes and nuclear atypia in ancient schwannomas may easily lead to a diagnosis of malignant mesenchymal neoplasm.[5] In our case, many of the schwannoma cell nuclei were large, hyperchromatic, pleomorphic and, often, multilobated. These nuclear features are known as degenerative nuclear atypia.[6] Absence of necrosis, mitosis, and invasiveness, as well as the absence of abrupt transition between typical schwannoma areas and cellular foci of atypical large cells, however, supported the diagnosis of ancient schwannoma.

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REFERENCES

1. Bayindir T, Kalcioglu MT, Kizilay A, Karadag N, Akarcay M. Ancient Schwannoma of the parotid gland. J Craniomaxillofac Surg 2006;34:38-42.
2. Ugokwe K, Nathoo N, Prayson R, Barnett GH. Trigeminal nerve schwannoma with ancient change. J Neurosurg 2005;102:1163-5.
3. Franz M. Enzinger, Sharon W. Weiss. Soft tissue tumor; 4th edition; 837.
4. Lanteri R, Rapisarda C, Santangelo M, Di Cataldo A, Licata A. Ancient schwannoma of the facial nerve. Chir Ital 2005;57:381-83.
5. Walther LE, Mentzel T, Sens A, Beleites E. Degenerative changes in parapharyngeal schwannoma "Ancient schwannoma". HNO 2001;49:54-8.
6. Nakayama H, Gobara R, Shimamoto F, Kajihara H. Ancient schwannoma of the oral floor and ventricular portion of the Tongue. Jpn J Clin Oncol 1996;26:185-8.