Case Report

Ancient schwannoma of neck masquedring as sarcoma

Tummidi Santosh, Sulata Choudhury[1], Prasanta Kumar Das[1], Manoj Kumar Patro[1]

Department of Pathology, Seth GSMC and KEMH, Mumbai, Maharastra, [1]Department of Pathology, MKCG Medical College, Berhampur, Odisha, India

ABSTRACT

Ancient schwannomas are rare benign encapsulated tumors of long standing duration. These tumors are usually solitary and may grow to a large size before detection of notable degenerative changes. The term "ancient schwannoma" is used to describe a schwannoma that has undergone changes such as relative loss of Antoni Type A tissue, perivascular hyalinization, calcification, cystic necrosis, hemorrhage and the presence of degenerative nuclear changes that may be misinterpreted as sarcomatous change. We report a case of ancient schwannoma in the left side neck.

Key words: Ancient schwannomas, fine needle aspiration, frozen section, histopathology, immunohistochemistry, sarcoma

INTRODUCTION

Ancient schwannoma, a degenerative neurilemmoma or schwannoma are characterized by, distinct degenerative morphological changes in the tumor that includes cystic necrosis, stromal edema, xanthomatous change, fibrosis, perivascular hyalinization, calcification, degenerative nuclei with pleomorphism, lobulation and hyperchromasia. These degenerative features are attributed to the growth and "aging" of the tumor, hence the term "Ancient schwannoma."[1] Growth of the tumor over time leads to vascular insufficiency, with resulting areas of tumor degeneration. Previous studies have correlated tumor size with progressive degenerative features. Despite these degenerative changes, ancient schwannoma behaves similarly to their conventional counterparts.[2] Schwannomas with these degenerative changes are often misdiagnosed as sarcoma or other soft-tissue neoplasms.[3] To the best of our knowledge, this is the first case report which provides a complete correlation between cytological, frozen section, histological and immunohistochemical features of ancient schwannoma.

Address for correspondence: Dr. Tummidi Santosh, Department of Pathology, Seth GSMC and KEMH, Mumbai, Maharastra, India.
E-mail: born_vss@yahoo.co.in

Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.200106

CASE REPORT

A 42-year-old male presented with a mass lesion on his left side of the neck. The swelling was slowly growing in size since last 8 years. The swelling was non-tender, mobile and the overlying skin was normal. Fine needle aspiration (FNA) showed pleomorphic spindle cells in loosely cohesive clusters as well as discretely over a proteinaceous and inflammatory background. Buckling of the nuclei was noted in some of these spindle cells. Mitotic figures were not seen. FNA impression was benign spindle cell lesion, possibly neural origin [Figure 1].

Following which intraoperative frozen section of the mass was advised to rule out the possibility of any sarcomatous component [Figure 2]. On gross examination, the specimen received in formalin was an encapsulated solid mass of 7 cm × 5 cm × 3 cm with attached adipose tissue. The tumor was soft to firm and well circumscribed. On sectioning, the

Cite this article as: Santosh T, Choudhury S, Das PK, Patro MK. Ancient schwannoma of neck masquedring as sarcoma. Clin Cancer Investig J 2016;5:537-9.

mass was tan in color, myxoid and soft in consistency and showed multiple foci of hemorrhage and necrosis [Figure 3]. Microscopic examination revealed an encapsulated tumor with solid cellular areas, hypocellular areas and a large cystic space. Cellular Antoni-A areas were interspersed with hypocellular Antoni-B areas surrounded by a fibrous capsule. The Antoni-A areas showed Verocay bodies. In Antoni-B areas cells were loosely arranged in a myxoid matrix, infiltrated by lymphocytes and pigment-laden macrophages along with multiple microcyst formations, hemorrhage and necrosis were seen. The Schwann cell nuclei were large, hyper chromatic and multilobulated. cytoplasmic vacoulations were seen. There were scattered ectatic irregular vessels with surrounding hyalinization. Differentiated Schwann cells immunohistochemically expressed S-100 protein in a diffuse and strong staining pattern [Figure 4]. Based on the above details, a diagnosis of ancient schwannoma was made. The patient was absolutely alright without any recurrence in the 5 months of the follow-up period.

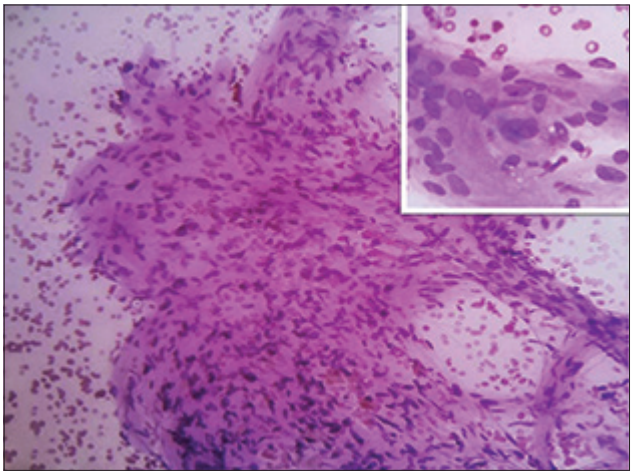

Figure 1: Cytology of lesion showing pleomorphic and bizarre cells, dark nuclei and some with vacuoles (inset), overall an appearance of benign spindle cell lesion (Diff-Quik, ×400)

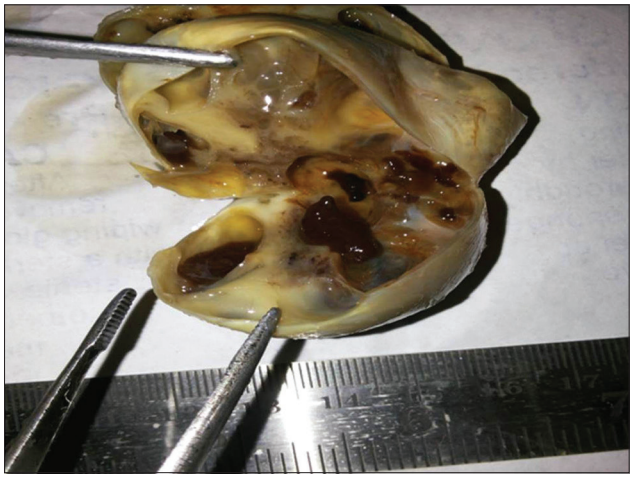

Figure 3: Gross specimen cut open shows a well-encapsulated tumor mass containing both solid and cystic areas with hemorrhage

DISCUSSION

Schwannoma also termed as neurilemmoma or neurinoma is one of the most common soft-tissue tumors. It is a benign tumor that arises from the Schwann cells of the nerve sheath and presents with symptoms of pain or paraesthesia. Tumor consists of two components: Antoni-A areas are more organized and are hypercellular, composed of spindle cells arranged in short bundles or interlacing fascicles. Antoni-B regions are hypocellular, less organized and contain more myxoid, loosely arranged tissue, with high water content. These components are intermixed within schwannomas and occur in varying amounts.[4]

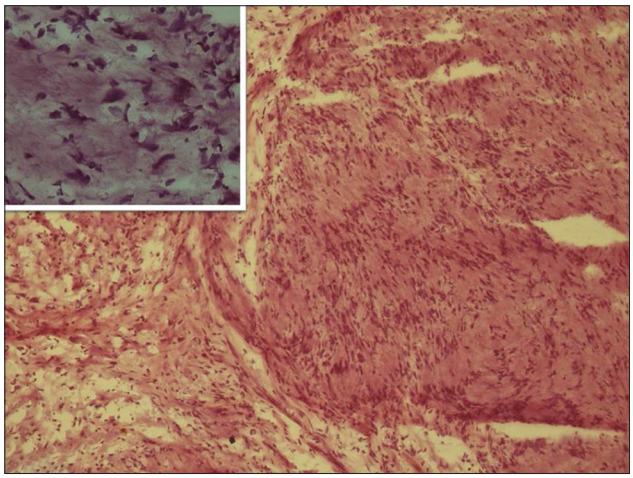

Figure 2: The frozen section from the tumor tissue showing spindle cell hypercellular areas (Antoni A) and hypocellular, myxoid areas (Antoni B). Degenerative nuclear changes can also be seen (H and E, ×400)

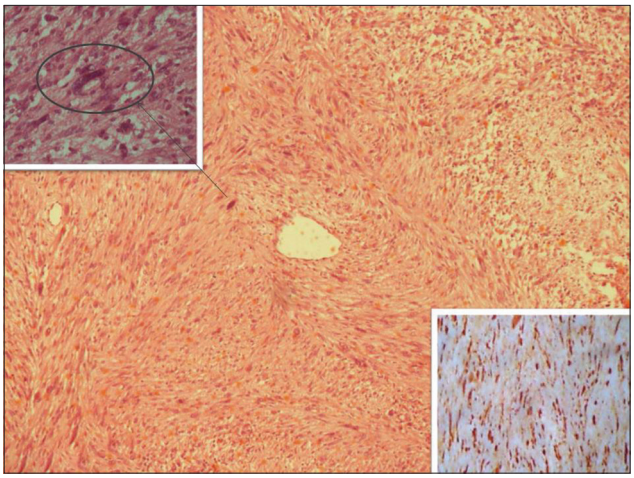

Figure 4: Routine microscopy revealed alternate hypercellular (Antoni A) and hypocellular (Antoni B) areas with areas of nuclear degenerative changes (inset A) and absent mitotic figures (H and E, ×400). Immunohistochemical staining of the tumor cells revealed diffuse, strongly positive staining for S-100 protein (inset B)

The term "ancient neurilemmoma" was first suggested by Ackerman and Taylor in a review of 48 neurogenic tumors of the thorax. The cited authors reported ten patients with tumors showing features similar to those of typical neurilemmomas, but differing in that significant tumor portions contained only a few cells within hyalinised matrices. They found that these features occurred in schwannomas of long duration, deeply situated, larger masses and hence coined the term "ancient schwannoma" representing 0.8% of all soft-tissue tumors.[5]

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.[6] A significant percentage of ancient schwannomas are located in deep locations such as the retroperitoneum. Ancient schwannomas of the neck are frequently misdiagnosed, and preoperative investigations are therefore often fruitless.[7] The histopathological features such as degenerative changes and nuclear atypia in ancient schwannomas may easily lead to a diagnosis of malignant mesenchymal neoplasm.[8] In the present case, many of the tumor cell showed large, hyperchromatic, pleomorphic, and often multilobated nuclei. These nuclear features are degenerative nuclear atypia. The absence of necrosis, mitosis, and invasive growth pattern 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. Immunohistochemistry for S-100 protein showed intense staining, suggesting a neural origin and is helpful in diagnosis, especially of a totally cystic degenerated mass.[8]

The treatment of choice for neck schwannoma is complete surgical resection of the tumor. Recurrence and malignant transformation of schwannomas are very rare. Malignant schwannomas are distinguished from benign schwannomas based on the presence of atypical mitosis and necrosis.[9]

CONCLUSION

Ancient schwannomas are slow growing, benign solitary neoplasm, rare in the neck region. Cytological findings were in correlation with long-standing nature of lesion. Complete excision of mass with intraoperative frozen section study followed by routine histopathological examination showed features of benign schwannoma. Surgery is the treatment of choice. The rate of recurrence and malignant transformation is very rare.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Dodd LG, Marom EM, Dash RC, Matthews MR, McLendon RE. Fine-needle aspiration cytology of "ancient" schwannoma. Diagn Cytopathol 1999;20:307-11.
2. Dahl I. Ancient neurilemmoma (schwannoma). Acta Pathol Microbiol Scand A 1977;85:812-8.
3. Kenichi I, Tominaga S, Tsutomu A, Hiroyuki K. Imaging of ancient schwannoma. AJR 2004;183:331-6.
4. Kransdorf MJ, Murphey MD, editors. Neurogenic tumors. Imaging of Soft Tissue Tumors. 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2006. p. 328-80.
5. Ackerman LV, Taylor FH. Neurogenous tumors within the thorax; A clinicopathological evaluation of forty-eight cases. Cancer 1951;4:669-91.
6. Bayindir T, Kalcioglu MT, Kizilay A, Karadag N, Akarcay M. Ancient schwannoma of the parotid gland: A case report and review of the literature. J Craniomaxillofac Surg 2006;34:38-42.
7. Bindra R, Gupta S, Gupta N, Asotra S, Sharma A. Ancient schwannoma of the neck mimicking soft tissue sarcoma. J Cancer Res Ther 2010;6:234-5.
8. Goldblum JR, Flope AL, Weiss SW. Benign tumors of peripheral nerves. In: Enzinger FM, Weiss SW, editors. Soft Tissue Tumors. 6th ed. Philadelphia, PA: Elsevier; 2014. p. 823-4.
9. Unni KK. Schwannoma. In: Fletcher CD, Unni KK, Fredrik M, editors. WHO Pathology and Genetics of Tumours of Soft Tissue and Bone. 4th ed. Lyon: IARC; 2002. p. 331.