

Case Report

Parachordoma: a rare recurring case at a rare site

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ABSTRACT

Parachordoma is an uncommon tumor of soft tissue and the origin is not clear. This soft tissue tumor resembles chordomas as well as extraskeletal myxoid chondrosarcomas and has only recently been fully characterized. Although it is considered a benign lesion, its behavior tends to be locally aggressive, with reports of a recurrence rate of up to 20% and of several cases of metastasis. In this article, we report a case of parachordoma in the neck with recurrence that we met in clinical works.

Keywords: Parachordoma, Neck, Recurrence

INTRODUCTION

Parachordomas are rare soft tissue tumors. They commonly occur in extremities, the lower being more common than the upper. Parachordomas also occur in chest, back, and abdomen. They have been included in 2002 world health organization (WHO) classification of soft tissue tumors under the category of “tumors of uncertain differentiation” along with mixed tumors and myoepithelioma. They are slow growing, benign lesions occurring at different ages. A small number of these tumors develop local recurrences and are rarely seen to metastasize.¹

We report a case of recurrent parachordoma in the neck with duration of 20 years.

CASE REPORT

A 65-year-old male presented with recurrent painless neck swelling of duration 20 years (Figure 1). On physical examination swelling measuring 3 cm by 2 cm was visible and palpable over the right aspect of the neck. The mass was soft, localized in the subcutaneous tissues, non-tender, and mobile. No imaging studies were

obtained, surgical resection was done. On gross examination specimen is cystic measuring 6 cm in largest diameter with focal dark brown areas.



Figure 1: Clinical photo.

Fine needle aspiration cytology was done and microscopy showed moderately cellular smears with abundant chondromyxoid background, admixed with tumor cells seen in cords, cluster & in singles, these cells are small

round to oval with vesicular chromatin, indistinct nucleoli, regular smooth nuclear membrane, cytoplasm is abundant, vacuolated & fragile (Figure 2).

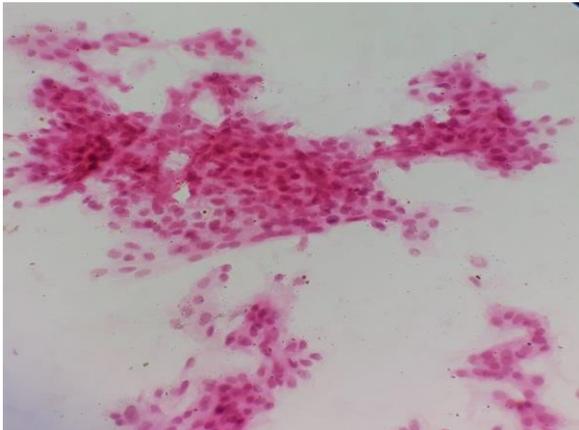


Figure 2: Cytology H&E (40x).

HPR showed small round to oval cells arranged in cords, chains and pseudoacini with abundant vacuolated cytoplasm against abundant chondromyxoid stroma (Figure 3).

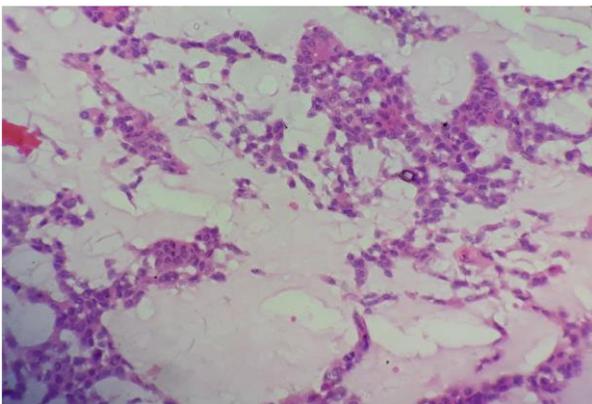


Figure 3: Histology H&E (40x).

On immunohistochemical testing it stained positive for CK 8/18 (Figure 4), S100 (Figure 5), and negative for EMA (Figure 6) and GFAP (Figure 7).

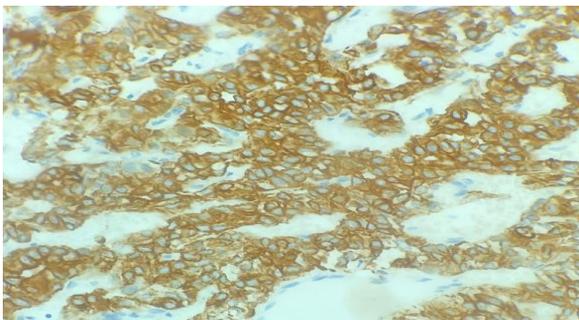


Figure 4: CK 8/18 - positive.

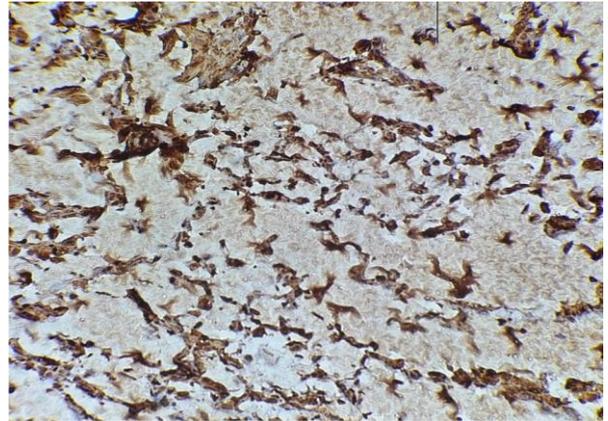


Figure 5: S-100 - positive.

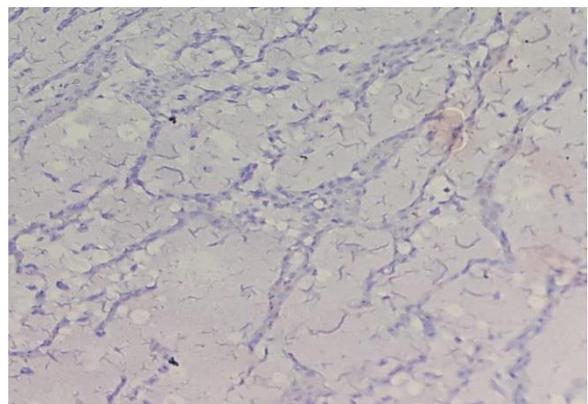


Figure 6: EMA - negative.

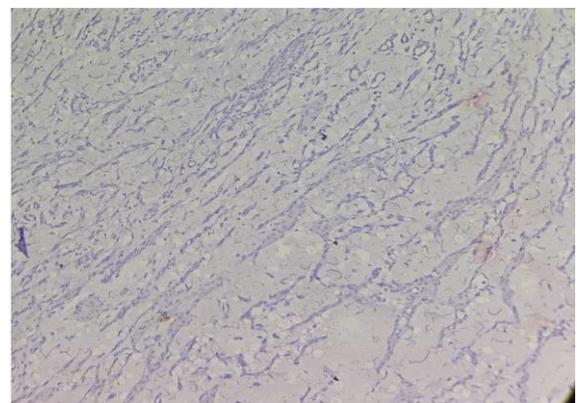


Figure 7: GFAP - negative.

DISCUSSION

Parachordoma is a kind of soft-tissue tumor whose origin is unknown. In 1977, Dabska reported five new cases and coined the term parachordoma. Laskowski first described this entity as extra-axial chordoma in 1995, with a series of 5 cases.¹ It is attributed to be ectopic rests of notochord, Schwann cells, myoepithelial cells or specialized synovial cells with no convincing evidence one way or another.²

Parachordomas are usually seen in adults but the 101-case series has reported an age range of 3 to 83 with a mean age of 39.4. The lesion is reported to be seen more commonly in males. The lower extremities are the most usual location.⁵ The location in the 101-case series was distributed as the lower extremity in 41, upper extremity in 35, head and neck in 15 and the body in 10.4. And in other study reported the most common location of parachordoma is the soft tissues of the extremities (78%), followed by the chest, the trunk, and the pelvis.² Parachordomas have been reported to occur in the pelvis and gastric serosa also.¹ Present case occurred in 65 year old male in the neck with history of recurrence twice.

Parachordomas clinically presents as a painless slow-growing nodular mass, which means that it usually goes unnoticed, and so diagnosis is normally established at a late stage.² However, two painful cases with a subperiosteal location have been reported.⁵ The tumor is well-limited and sometimes surrounded by a thin fibrous pseudocapsule. It is usually 3 to 7 mm in size and can grow up to 12 cm. present case presented as a painless swelling measuring 3×2×2 cm in size.

Parachordoma was initially considered to be a chordoma in nonaxial location, but now it is considered as a unique entity with distinct immunohistochemical profile. Parachordoma is considered a slow growing tumor of an indolent, less aggressive nature than chordoma. The differential diagnosis for this tumor includes extraskelatal myxoid chondrosarcoma, chordoid meningioma, and chordoma.¹ Immunohistochemistry is essential to differentiate this tumor from extraskelatal myxoid chondrosarcoma and chordoma. In the former, the cells are usually smaller and they do not stain with cytokeratin (CK). As for the latter, both tumors stain for S100 protein and epithelial membrane antigen but only chordoma expresses the t-box transcription factor brachyury.³

Immunohistochemistry for a variety of cytokeratins (CKs) (8/18, 1/10, 7, and 20), epithelial membrane antigen (EMA), S-100 protein, vimentin CD-34, type IV collagen, smooth muscle actin, smooth muscle myosin heavy chain, calponin, and glial fibrillary acid protein was performed in a study of six cases. All parachordomas strongly expressed CK 8/18, but not the other cytokeratins. Additionally, they expressed EMA (five of six), S-100 protein (six of six), and vimentin (six of six) and had a linear pattern of type IV collagen immunoreactivity around nests of cells (four of five). Calponin was noted in one case, but no cases expressed smooth muscle actin, smooth muscle myosin heavy chain, or glial fibrillary acid protein.⁶

More than 95% of parachordomas stain positive for CK, vimentin, and S100 protein.³ Present case stained positive for CK 8/18 and S-100 and negative for EMA and GFAP.

Parachordomas are benign tumors but metastases and recurrences are not unusual.² Dabska has reported recurrences after 7, 2 and 12 years later but Niezabitowski et al have reported a recurrence at 3 months, Carstens et al at 6 months and Ishida et al at 1 year although early recurrence is rare. However, it is difficult to determine the mean rate of recurrence due to the difficulty of a long-time follow-up.⁵ Our case had a recurrence after 6 years at the same site. Two deaths from metastatic parachordoma have been reported in the literature and it has been said that parachordomas may potentially be low-grade sarcomas. Parachordomas are benign tumors but they can become malignant when recurrence occurs after a short period and cases should therefore be followed-up closely.⁵

CONCLUSION

Parachordoma is a rare soft-tissue tumor with an unknown true incidence. Nevertheless, it should be considered in the differential diagnosis of soft-tissue masses, especially in the extremities of young adults. This tumor is recognized for its late recurrences. Thus, long term follow-up may be needed after the surgical resection even when tumor-free margins are achieved given recurrences can present more than 5 years after surgery.

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Ethical approval: Not required

REFERENCES

1. Ghanta RK, Uppin MS, Koti K, Hui M, Uppin SG, Mukherjee KK. Primary intracranial Parachordoma: An unusual tumor in brain. *Surg Neurol Int.* 2014;5:S506-11.
2. Estrems Díaz V, Bertó Martí FX, Zarzuela Sánchez V, Cabanes Ferrer MI, Bru Pomer A. Parachordoma of soft tissues of the arm: a very rare tumour. *Cas Repo Orthoped.* 2013;2013.
3. Belzarena AC, Mankanji RJ, Joyce DM. Recurrent parachordoma of the lower back: A case report. *Radiol Cas Repo.* 2019;14(1):94-6.
4. Zhang J, Wang H, Cheng X, Wang M, Zhu Y. A case of parachordoma on the chest wall and literature review. *J Can Res Ther.* 2013;9:114-7.
5. Karakaya YA, Ozekinci S, Büyükbayram H, Mizrak B. Parachordoma: a recurrent case and review of the literature. *Turk Patoloji Derg.* 2011;27:173-6.
6. Folpe AL, Agoff SN, Willis J, Weiss SW. Parachordoma is immunohistochemically and cytogenetically distinct from axial chordoma and extraskelatal myxoid chondrosarcoma. *Am J Surg Pathol.* 1999;23(9):1059.

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