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Case Report

Unveiling the mimic: Chondroid syringoma clinically masquerading as liposarcoma – A rare encounter

Archi Arun Chawla[1], Pradnya Paresh Chalke[1], Reeta Dhar[1], Shilpi Sahu[1], Kaivalya Bhaskar Shah[1]*

[1] *Dept. of Pathology, MGM Medical College, Navi Mumbai, Navi Mumbai, India*

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ABSTRACT

Chondroid Syringoma is an uncommon benign adnexal tumor of the skin, representing approximately 1% of frequently diagnosed skin tumors. This tumor consists of both epithelial and mesenchymal stromal elements, thus classified as a mixed skin tumor. Despite being primarily a benign sweat-gland tumor, chondroid syringoma typically manifests as a painless, gradually enlarging mass, commonly found on the head and neck area. In this particular instance, the tumor was situated on the lateral aspect of the right thigh, which is an unusual site for its presentation.

1. Introduction

Chondroid syringoma is a sporadic skin appendage tumor with an incidence of <0.01 to 0.098 percent.[1,2] Chondroid syringomas are rare, benign tumors that arise from sweat glands. These tumors exhibit a unique combination of epithelial and mesenchymal tissue components. Although chondroid syringomas typically develop in the head and neck region, the occurrence of these growths on the outer right thigh, as observed in this case, is exceptionally uncommon.[3] The initial description of this condition was made by Billroth, who referred to it as a "cutaneous mixed tumor."[1] Virchow and Minssen classified these tumors as mixed tumors. Chondroid syringomas typically manifest in individuals aged 20 to 60 years, with a higher incidence among men at a 2:1 ratio. The term "syringoma" refers to the consistent presence of exocrine gland elements, particularly those resembling sweat glands, within the tumor structure. The term "chondroid" was added to highlight the notable presence of cartilaginous or cartilage-like material in the tumor.[2,4,5] Chondroid syringomas typically present as slow-growing, painless, rounded, firm, elevated mobile masses beneath the skin. These features often lead to confusion with epidermal cysts, as both can share similar characteristics.[6]

Chondroid syringomas are typically benign, but there are rare instances of malignant cases reported in the literature. Malignant chondroid syringomas can exhibit features such as local recurrence, metastasis, and in severe cases, even death. Lesions with the size >9cm have a higher tendency to turn malignant. Lesions on the trunk and extremities have a higher chance of turning malignant, while benign ones are commonly found in the head and neck region.[1] The definitive diagnosis of chondroid syringoma can only be established through a histopathological examination of a complete specimen obtained through biopsy or surgical excision. Once diagnosed, the best treatment for chondroid syringoma is complete surgical excision along with adequate margins to ensure the removal of all tumor cells and reduce the risk of recurrence.[1,7,8]

* *Corresponding author*.
E-mail address: kaivalyashah7700@gmail.com (K. B. Shah).

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2. Case Report

A 38-year-old woman presented with a swelling in the region of the right thigh for past few years. The patient experienced intermittent episodes of pain that gradually intensified over time. She reported no prior medical conditions or family history relevant to the issue and described an intermittent white discharge from the affected area.

The Ultrasonography of right thigh swelling revealed a 6 x 5.5 x 4 cm heterogeneously iso to hyperechoic mildly lobulated solid lesion having a hypoechoic capsule, showing mild internal vascularity and posterior acoustic enhancement and calcifications suggesting it as

Pilomatricoma**.** MRI studies revealed a well-defined encapsulated 7.6 x 5.7 x 5.2 cm sized lobulated altered signal intensity lesion noted in the subcutaneous plane of the anterolateral aspect of the right thigh.

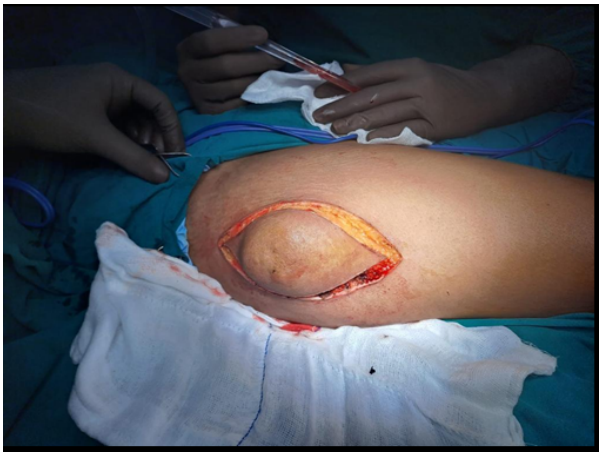

Figure 1: Gross Intra-operative image of the swelling over the Right Thigh.

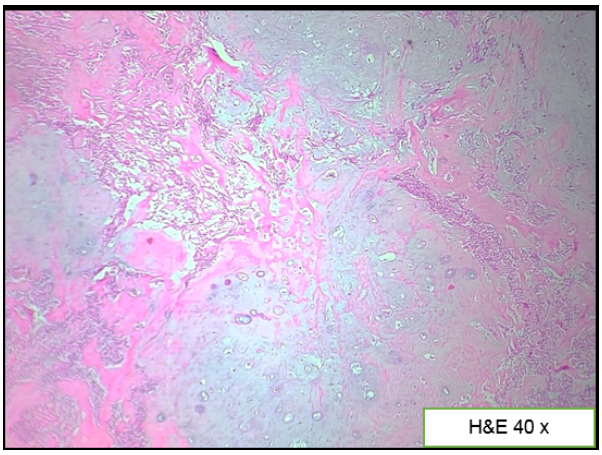

Figure 2: The image shows predominantly chondroid and myxoid stroma with little epithelial component

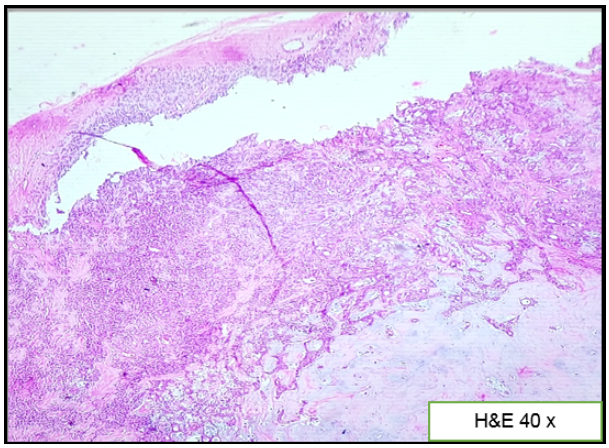

Figure 3: The tumour shows predominantly epithelial component in cord, nests, tubules along with chondroid focus at the right side.

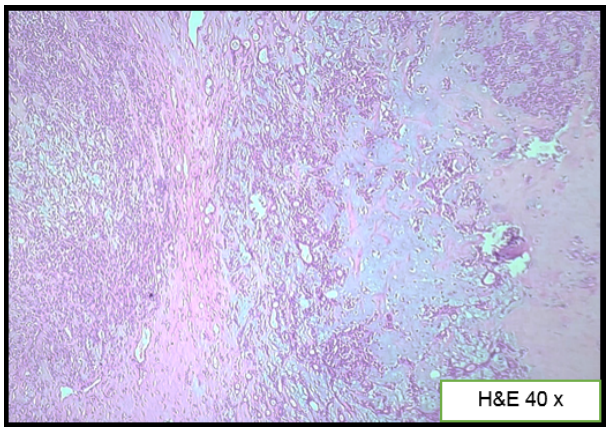

Figure 4: The tumor shows predominantly epithelial component in cord, nests, tubules along with chondro-myxoid focus at the right side.

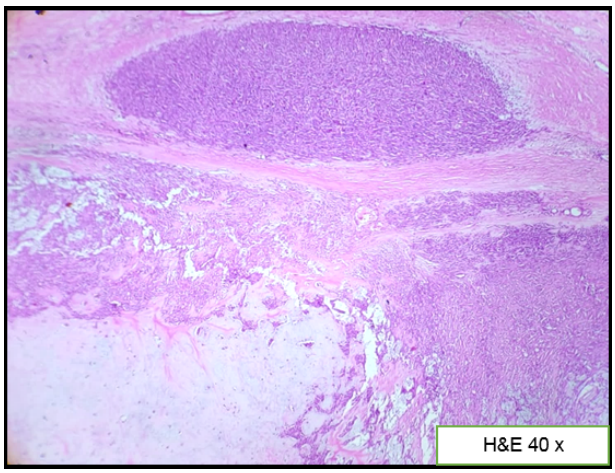

Figure 5: The tumour shows islands of benign epithelial cells amidst chondromyxoid, fibrous, and hyaline stroma.

We received the specimen in the Department of Pathology for histopathological examination.

On Gross examination, a single, grey-white to grey-brown, soft to firm skin-covered mass measuring 13 x 5 x 3cm with attached fibrofatty tissue was identified. The cut surface exposed a well-defined, round, uniform grey-white mass measuring 5 x 4.5 x 3.8 cm. The deep surgical margin was 1 cm from the edge of the mass.(Figure 1)

3. Discussion

Chondroid syringoma is occasionally referred to as a mixed tumor of the skin, reflecting its mixed epithelial and mesenchymal composition. The term "mixed tumor" was first introduced by Billroth in 1859 when he identified these lesions as benign mixed tumors of the skin. This historical recognition highlights the long-standing recognition and study of this uncommon yet distinctive skin tumor.[7]

In 1961, Hirsch and Helwig coined the term "chondroid syringoma" to distinctly identify tumors originating from sweat glands. They proposed that chondroid syringomas originate from either apocrine or eccrine sweat glands and emphasized their histopathological differences from pleomorphic adenomas, which arise from salivary glands. This designation helped to further clarify the classification and understanding of these unique skin tumors.[9] Chondroid syringomas are regarded as exceptionally rare, with an incidence ranging between 0.01% to 0.098%. They typically exhibit a male predominance and are most commonly observed in individuals aged between 20 and 40 years. While they predominantly appear on the head and neck, cases have been reported in other regions of the body as well. This rarity underscores the importance of recognizing and properly diagnosing these tumors when they do occur.[3,4] The nodules are usually light red or skin-colored and have smooth surfaces. Medium-hard or hard texture nodules are thought to be a result of their interstitial component. Most masses have diameters of 0.5 and 3 cm; tumors as large as 9 cm have also been documented. In our patient, the nodule was at an uncommon location with a large size of 13 x 5 x 3cms[1]

Hirsch and Helwig named it chondroid syringoma because of the presence of sweat gland elements in a cartilaginous stroma which is the histological criteria for diagnosis.

The histological features observed include nests of cuboidal or polygonal cells and intercommunicating tubuloalveolar structures lined with two or more rows of cuboidal cells (Figures 2 and 3). Ductal structures are composed of one or two rows of cuboidal cells, and occasional keratinous cysts are present. Additionally, there is a varying composition of chondroid and myxoid matrix (Figures 4 and 5). Chondroid syringomas may have all five characteristics or manifest only some.

In our case all the above features are present. Chondroid syringoma is an uncommon lesion having male predominance usually in middle or old age.[7]

Histopathology indeed serves as the gold standard for diagnosing chondroid syringomas showing two main types which can be identified as 1) tubular and cystic lumina that partially branch, and 2) smaller tubular lumina. This more common type exhibits significant variations in the size and shape of its tubular lumina, including cystic dilatation and branching. The tubular lumina are lined by two layers of epithelial cells within a dense stroma - a luminal layer of cuboidal cells and a peripheral layer of flattened cells. This histological pattern aids in distinguishing chondroid syringomas from other skin lesions.[10]

Surgical excision is surely the most effective treatment for Chondroid Syringoma, removing the surrounding capsule entirely to reduce the risk of recurrence. Periodic follow-ups are certainly useful.[5]

4. Conclusion

The presence of a slow-growing hard nodular lesion in a middle-aged patient should raise suspicion for chondroid syringoma among other differential diagnoses. Accurate diagnosis relies on comprehensive history-taking and histopathologic examination. Surgical excision remains the standard treatment of choice. However, it is crucial to exercise caution during excision to ensure complete removal of the tumor mass and minimize the risk of recurrence. Long-term follow-up is strongly recommended to facilitate early detection of any recurrence and to manage it promptly also to minimize the likelihood of complications associated with recurrence.

5. Source of Funding

None.

6. Conflict of Interest

None.

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Author biography

Archi Arun Chawla, Junior Resident

Pradnya Paresh Chalke, Junior Resident

Reeta Dhar, Professor

Shilpi Sahu, Professor and HOD

Kaivalya Bhaskar Shah, Junior Resident

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