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Case Report

Large benign chondroid syringoma of the lower limb-an uncommon presentation of a rare disease

Ashwin Vinod, George Mathews John*, Ranjith V. B., Devanarayan V.

Department of General Surgery, Amrita Institute of Medical Sciences, Amrita Vishwa Vidyapeetham, Kochi, Kerala, India

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***Correspondence:**
Dr. George Mathews John,
E-mail: gmjohn07@gmail.com

ABSTRACT

Chondroid syringoma (CS) is a sporadic skin appendage tumor that manifests as slow-growing, subcutaneous, non-tender, non-ulcerating dermal nodules, most often in the head and neck. The tumor is more common in men, has a biphasic age distribution, and usually attains a size of 0.5 to 3 cm. Larger lesions up to 9 cm in size have been recorded in the literature, with the majority of them containing malignancies. Rare cases of malignant CS have been described, with local recurrence and metastasis. We report a case of an 85-year-old man presenting with complaints of a significantly large swelling in the inner aspect of his right thigh for the past 25 years. Clinical and radiological differential diagnoses of a large uncomplicated dermoid cyst, sebaceous cyst, epidermal inclusion cysts, neurogenic tumours, or myxoid tumours with cystic degeneration were suggested. Under general anaesthesia, the lesion was completely excised with a 1 cm margin of healthy surrounding tissue. The histopathological examination of the complete surgical specimen was consistent with CS. Patient has been on regular follow-up and has shown excellent response to treatment, with no evidence of recurrence, metastasis, or disfigurement. With this paper, we report a case of benign CS in an unusual location, attaining a huge size (>10 cm) and, even then, not turning malignant. We hope to add to the current knowledge on CS, allowing for early and accurate diagnosis and successful disease management.

Keywords: Benign or malignant skin conditions, CS, Mixed tumor of skin, Skin appendage tumor

INTRODUCTION

Chondroid syringoma (CS) is a sporadic skin appendage tumor with an incidence of less than 0.01 to 0.098 percent.[1,2] The disease was first described by Billroth as a "cutaneous mixed tumor' and later revised by Hirsch et al., who coined the term ' CS ' in 1961.[3] It arises from the sweat glands and is similar to the pleomorphic adenoma of the salivary glands. The tumor is more common in men and has a biphasic age distribution.[1,3]

CS manifests as slow-growing, subcutaneous, non-tender, non-ulcerating dermal nodules, most often in the head and neck.[1,3,4] The tumor has epithelial and mesenchymal components and sweat-gland parts in a cartilaginous stroma. CS is often benign, although rare cases of malignant CS have been described, with local recurrence, metastasis, and death.[5-7] Larger lesions up to 9 cm in size have been recorded in the literature, with the majority of them containing malignancies.[6,7] Malignant CS is frequently seen on the trunk and extremities, whereas benign CS is found in the head and neck.[7]

The condition's rarity can be easily misdiagnosed with other cutaneous lesions like a sebaceous cyst, dermoid cyst, and basal cell carcinoma. Fine needle aspiration cytology samples from the lesion may provide some information about nature of tumor. However, definitive diagnosis can be made only by histo-pathological examination of complete specimen. Histopathological

examination usually reveals a nest of cells and lace-like pattern of strands of epithelial cells and tubuloalveolar structures, occasionally keratinous cysts, and varying amounts of the matrix.[1,3,6] The optimal treatment for CS is complete excision with adequate margins.[1,2,7]

Benign CS is extremely rare, with very few previously reported cases, all of which are less than 5 cm in size and mainly in head and neck region.[1-5] Large lesions reported malignant and up to a size of 9 cm in largest diameter.[6,7] We report case of benign CS in an unusual location, attaining a huge size and even then not turning malignant.

CASE REPORT

An 85year-old man presented to our hospital with complaints of a significant swelling in the inner aspect of his right thigh for the past 25 years. The swelling was initially small in size and showed a recent increase in size, causing hindrance to the usual walking of the patient. His medical background was significant for systemic hypertension, coronary artery disease, and late-onset bronchial asthma, all controlled with medications. On clinical examination, a smooth, non-tender, non-fluctuant, subcutaneous swelling with variegated consistency measuring 12×10 cm was found in the posteromedial aspect of the upper one-third of the right thigh (Figure 1A). The swelling showed partial fixity to the overlying skin and no fixity to the underlying muscle. No similar swellings were noted elsewhere on the body. Clinical differential diagnoses of a large uncomplicated dermoid cyst, a sebaceous cyst, or a cutaneous soft tissue tumor were made.

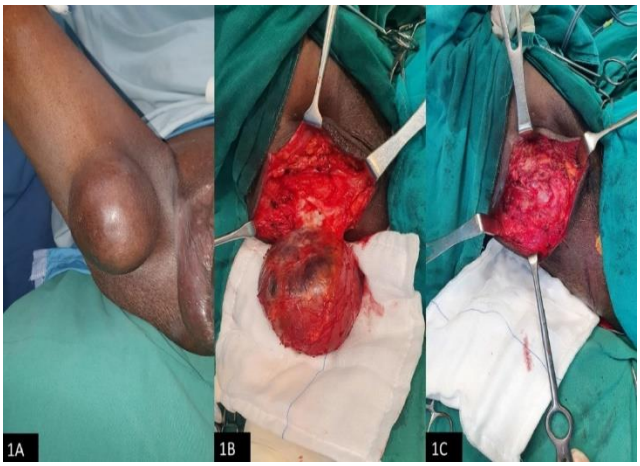

Figure 1 (A-C): Preoperative and postoperative images of swelling. Clinical presentation of smooth, round-to-oval, non-tender, non-fluctuant subcutaneous swelling in upper 1/3rd right thigh, measuring approximately 12×10 cm. Surgical specimen *in situ* after almost complete dissection-swelling with variegated consistency and significant vascularity in subcutaneous plane. Tumor bed after removal of lesion in toto-showing normal surrounding tissue and no infiltration into underlying muscle.

A contrast-enhanced computed tomography of the abdomen and pelvis showed a thin-walled, well-encapsulated, oval cystic mass in the right upper thigh region measuring 10.8×8.8×8.1 cm (Figures 2A and B). The soft tissue mass was in the subcutaneous plane and had no deeper extension. No calcifications, no air-fluid levels, and no fat density or solid enhancing areas were noted within the lesion. Radiological differential diagnoses of benign lesions like epidermal inclusion cysts, neurogenic tumors, or myxoid tumors with cystic degeneration were suggested.

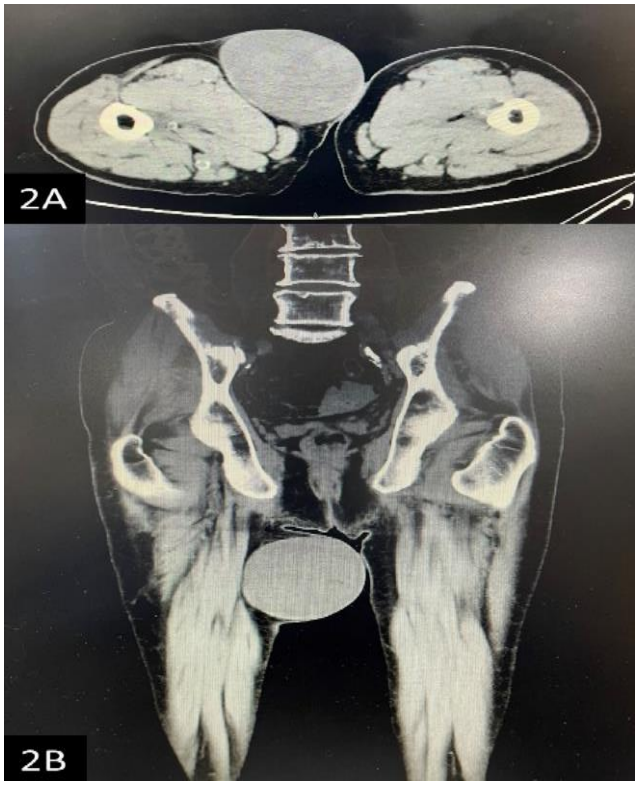

Figure 2 (A and B): Contrast-enhanced computed tomography images of the swelling, axial section; coronal section; CECT of the abdomen and pelvis showing a thin-walled, well-encapsulated, oval cystic mass in the right upper thigh region measuring 10.8×8.8×8.1 cm.

Under general anesthesia, the lesion was completely excised with a 1cm margin of healthy surrounding tissue (Figures 1 B and C). A part of the overlying skin was also excised along with the specimen. The specimen measured 10.5×8.1×8.2 cm (Figures 3A and B). The surgical wound was closed in layers following surgical principles. A surgical drain was placed in situ before the closure of the wound. The wound was healthy, and the surgical drain was removed on the third postoperative day. A two-week follow-up consultation in the outpatient clinic showed a completely healed wound with no deformity. The patient was kept on a once-in-two-week follow-up for six weeks and showed excellent clinical response to treatment, with the wound ultimately healing, a healthy scar, and no signs of any complications.

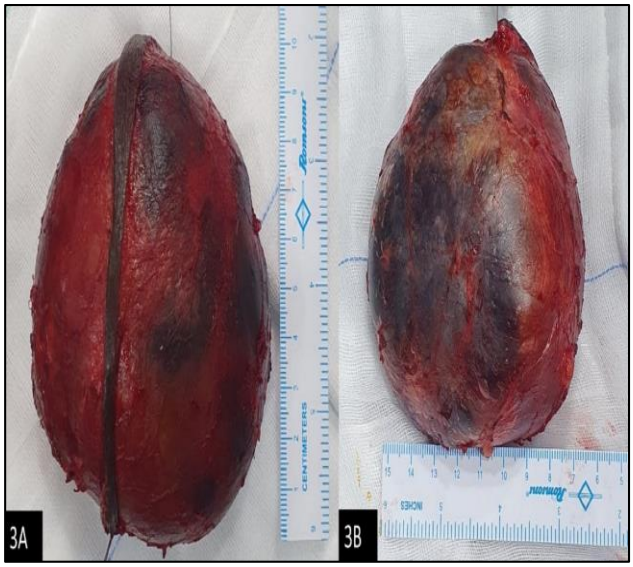

Figure 3 (A-B): Gross pathology images of the surgical specimen, anterior view showing the skin bridge excised along with the specimen as well as variegated consistency and posterior view showing variegated consistency of the specimen.

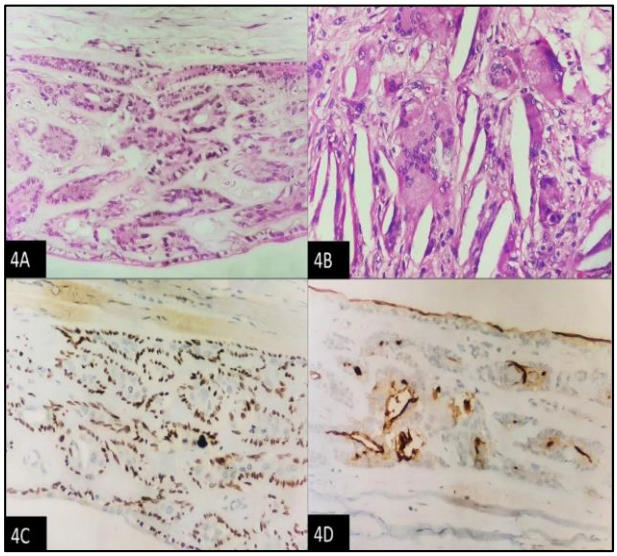

Figure 4: Histopathology and immunohistochemistry images of the specimen, tubules lined by two layer of cells, outer myoepithelial cells with clear cytoplasm and inner luminal cells (400x). The surrounding stroma showing basement membrane-like material intermixed with chondromyxoid stroma; foreign body giant cell reaction in cyst wall (400x). P63 positivity highlighting outer myoepithelial cells (IHC 400x). EMA showing positivity in luminal cells (IHC 400x).

The specimen measured 10.5×8.1×8.2 cm.

Routine histopathological analysis of the excised specimen showed the tumor lumen containing abundant fibrinous material. The luminal surface of the cyst was lined by cuboidal cells having eosinophilic cytoplasm and apical snouts. Cells arranged as bilayer tubular patterns containing eosinophilic secretion were also noted. The surrounding stroma showed basement membrane-like material intermixed with chondromyxoid stroma (Figures 4 A and B). Immunohistochemical analysis of the specimen showed p63 positive in the outer layer (myoepithelial cells) and EMA positive in the luminal and cyst lining cells (Figures 4 C and D). Based on the morphology and immunohistochemistry findings, a final diagnosis of benign appendageal neoplasm with apocrine and eccrine differentiation suggestive of a CS was made.

No evidence of any deformity or local recurrence was noted at 12 weeks and 16 weeks follow-up visits. The patient was delighted with the surgical outcome and noticed a significant improvement in his ambulation and ease of carrying out his daily routines. Due to the long duration of the disease at the time of presentation, close resection margins, and large lesion size, the patient has been advised regular clinical follow-up.

DISCUSSION

The uncommon and mostly benign tumor of the skin, known as CS, is sometimes referred to as a mixed tumor of the skin (MTS).[3] According to the literature, the incidence of CS accounts for as little as 0.01% to 0.098% of primary skin cancers [1,3]. Middle-aged males are most commonly affected by CS, which primarily affects the head and neck.[1,6,7] A solitary subcutaneous nodule with a modest development and a distinct border is one of the clinical characteristics of CS. The nodules are usually light red or skin-colored and have smooth surfaces. The nodules' medium-hard or hard texture is thought to be a result of their interstitial component.[1,6,7] Most masses have diameters between 0.5 and 3 cm; however, tumors as large as 9 cm have been documented.[1-7] The nodules of CS typically do not ulcerate even on attaining a large size. In our patient, the nodule was at an uncommon location and attained a large size of 12×10 cm.

The histological manifestations of CS can take many different forms, although they often present as epithelial components embedded in a matrix that resembles mucus, cartilage, or fibrous tissue.[8] Apocrine and eccrine CS are two subtypes of CS.[8-10] Apocrine CS exhibit a comparatively wide tubule lumen under the microscope. There may be apocrine differentiation as evidenced by the two layers of cells covering the lumen wall, branch ducts, and the presence of apocrine secretion.[9] Eccrine secretion and relatively tiny, circular, branchless gland ducts make up eccrine CS. In addition to the two adenoid forms mentioned above, there are additional epithelial bands, islands, and solitary epithelial cells.[8] In some circumstances, sebaceous glands or visible hair germ cells occur together with hair follicle development. The majority of these instances include apocrine CS. The routine histopathological examination of the surgical specimen in our case showed a combination of apocrine and eccrine differentiation consistent with CS.

In certain instances, nodules resembling pilomatrixoma are formed due to giant-cell foreign-body response, eosinophilic ghost cells, and hair matrix differentiation.[9,11] Cell-rich CS are cases that have a high concentration of cellular components (>95%) but no cartilage interstitium or mucus.[12] CS has occasionally invaded small blood vessels, but there has been no recurrence or metastasis over several years of follow-up. Hyalinization may occur as a consequence of CS epithelial cells. In around 40% of CS, glass-like cells can be seen. These cells are classified as plasmacytoid cells because they resemble plasma cells and have an oval shape and intensely colored cytoplasm that resembles glass and deviated nuclei. These cells should be referred to as "plasmacytoid myoepithelial cells" because the literature indicates they possess myoepithelial characteristics.[13] These cells typically exhibit two immunohistochemical expression patterns since they are still in the pluripotent stage, during which it is difficult to establish the direction of differentiation or maturation.[14]

Immunohistochemistry findings have been reported in cases of CS, including positivity towards CK, CEA, EMA, vimentin, S-100, GFAP, p63, SMA, and many more.[6,15] The immunohistochemical staining in our case showed p63 and EMA positivity, which are sensitive but non-specific markers. Hence, hematoxylin and eosin staining should be used to determine the accurate diagnosis, as immunohistochemistry of CS lacks specificity.

Clinical signs of CS are also not very precise. Clinical testing should distinguish CS from other cancers, such as Syringoma, dermatofibroma, trichoepithelioma, and epidermal cysts.[16] However, a pathological investigation is crucial to confirm the diagnosis of CS. The differential histological diagnosis should distinguish it from benign adnexal tumors such as apocrine adenoma, pilomatrixoma, clear-cell hidradenoma, and sebaceous cysts.[17] It should be highlighted that the microscopic appearance of MTS is quite similar to the pleomorphic adenoma from the salivary gland, making it a challenge to differentiate.

Since CS is usually a benign lesion, it may be removed surgically without the risk of local recurrence. Although several treatments for this tumor have been suggested, such as electrodesiccation, dermabrasion, and laser vaporization using argon or CO2, total surgical removal is the preferred method.[1,3,7] Long-term follow-up is not indicated if the tumor has been excised and is benign.[1] For the patient discussed in this article, we achieved complete surgical excision, and the patient showed satisfactory clinical and pathological response to the treatment. No additional treatment procedures were required.

Nevertheless, a possibility of malignant CS exists, according to the literature, considering the enormous size of the lesion. Malignant CS exhibits heteromorphic cell expansion, a rise in nuclear fission, infiltrative growth of the margin, satellite lesions, and tumor necrosis at the microscopic level.[1,3] These features were absent in our case, giving it a <5% risk of malignant transformation. Unconfirmed reports show that the dura is affected by a rare kind of malignant CS in the scalp region.[17-19] Lymph nodes, the lung, the bone, and even the central nervous system can all experience tumor metastasis.[1,3,7,18,19] Therefore, we recommend establishing a follow-up protocol following the complete excision of malignant CS. For malignant CS, the treatment of choice is wide excision and adjuvant radiation therapy with or without chemotherapy.[18,19] Malignant CS seldom recurs following resection, and in such cases, we recommend micrographically-oriented-histographic-surgery (MOHS) to remove the malignant CS.[20] The patient mentioned in this article is still on regular follow-up and has shown no evidence of any recurrence or distant metastasis.

CONCLUSION

CS is rare, and a surgeon confronted with such a tumor has a clinical dilemma. A middle-aged patient with a slow-growing nodular lesion should alert the surgeon to the likelihood of a CS. Accurate history-taking and histopathologic testing are required for diagnosis. The standard treatment of choice is surgical excision. A word of caution should be issued regarding the complete excision of the tumor mass since it may recur later. A long-term follow-up is suggested for early detection and to reduce the possibility of recurrence.

With this report, we aim to enhance the existing literature on CS, thus enabling early and accurate diagnosis and effective management of this rare surgical entity. We also aim to highlight the importance of histopathological diagnosis in a case of CS, as clinical symptoms, radiological features, and immunohistochemistry lack specificity.

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