Clinical Case Report

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A rare large cutaneous chondroid syringoma involving a toe

A case report

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Abstract

Rationale: Chondroid syringoma (CS) occurs mostly on the face and neck, and rarely occurs in the toe. Malignant CS is invasive, grows quickly, and has a high recurrence rate. The presence of a bilobed CS in 1 toe has never been reported in the literature.

Patient concerns: A 72-year-old male patient presented with a mass in a third toe of his right foot. The mass had slowly grown in 2 years. He felt mild pain and the mass occupied most of the tip of the toe.

Diagnoses: Radiographs showed a large soft-tissue mass in the third toe of his right foot without any bone destruction. Ultrasonogram showed 2 partly fused hypoechoic masses within the lesion. The mass was therefore diagnosed as a benign CS.

Interventions: We amputated the toe with the mass under local anesthesia. The postoperative pathohistological examinations confirmed that the lesion was a bipartite CS exhibiting active cellular proliferation.

Outcomes: Two years after surgery, there was no tumor recurrence.

Lessons: CS can also present as multiple adjacent masses. Complete surgical resection and long-term follow-up are essential.

Abbreviations: AFP = alpha-fetal protein, CA19-9 = cancer antigen 19-9, CEA = carcinoembryonic antigen, CS = chondroid syringoma, EMA = epithelial membrane antigen, G15 = gross cystic disease fluid protein 15, HE = hematoxylin and eosin, MMG = mammaglobin, PSA = prostate-specific antigen, SMA = smooth muscle actin.

Keywords: chondroid syringoma, surgical resection, toe

1. Introduction

Chondroid syringoma (CS) is a type of rare skin tumor that is often benign. It usually occurs on the head and neck.[1] Malignant CS and malignant transformation of a benign CS is possible but uncommon.[2] Treatment usually consists of surgical excision. We present here a case of a bilobed CS on 1 toe, which has never been reported in the literature.

2. Case presentation

A 72-year-old male patient presented with a mass on his third toe of his right foot and the mass had slowly increased in size in 2 years (Fig. 1). The mass caused mild pain, and the patient reported no history of trauma of the toes of his right foot. On local examination, the mass was firm and located on the volar side of the toe tip. No abnormalities were found during the

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Ethics approval: These study protocols were approved by the Medical Ethics Committee of the First Affiliated Hospital, College of Medicine, Zhejiang University.

Consent for publication: Written informed consent for publication of clinical details and images were obtained from the patient. A copy of the consent form is available for review upon request.

Availability of data and materials section: The data supporting the conclusions of this article are included within the article.

Competing Interests

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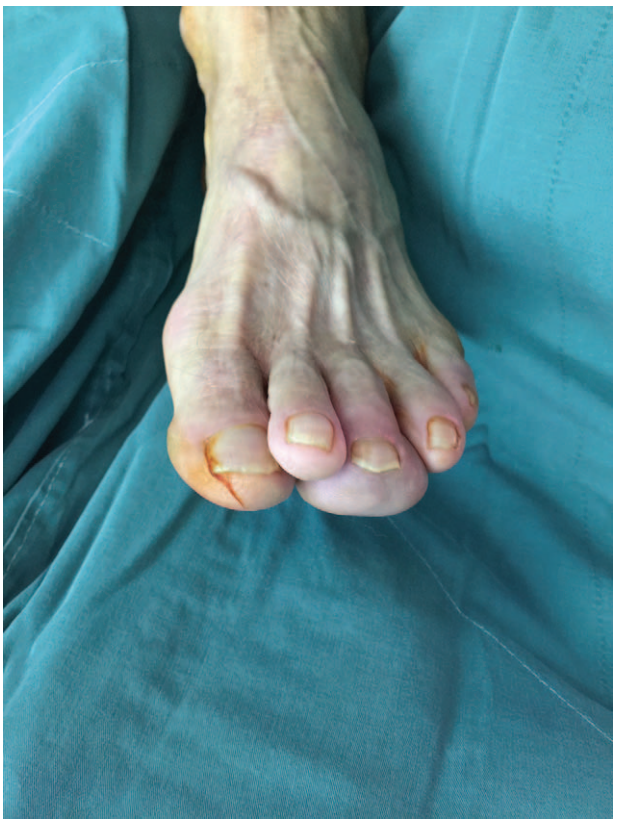

Figure 1. Preoperative photograph showing a large tumor in the third toe of the right foot.

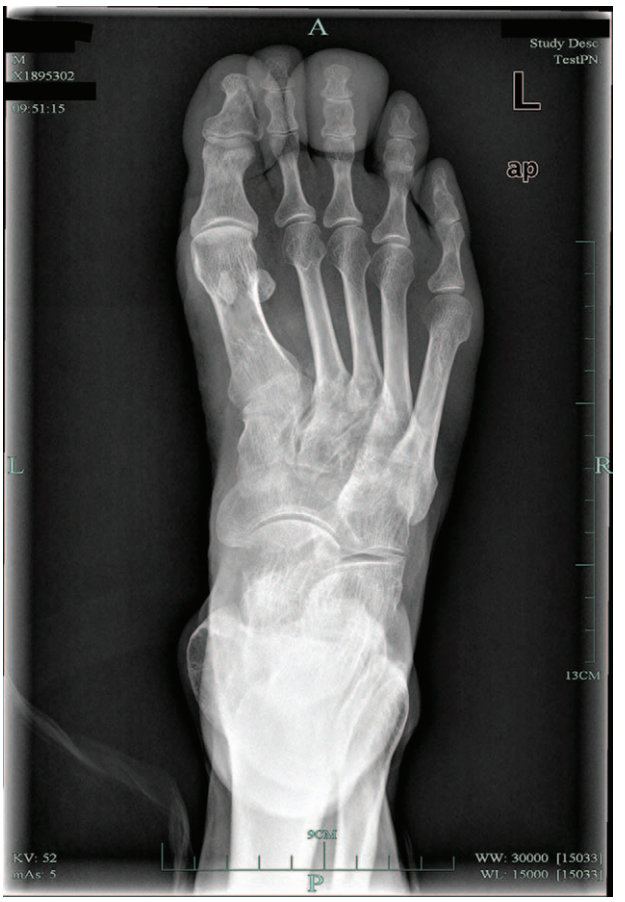

Figure 2. Radiograph showing a large soft-tissue mass in the third toe of the right foot without any bone destruction.

inguinal, liver, and lung imaging explorations. Neurological examination, tumor biological markers (alpha-fetal protein, prostate-specific antigen, carcinoembryonic antigen, cancer antigen 19-9), and other laboratory tests including complete blood count and electrolytes, were all normal. Radiographs showed a large soft-tissue mass in the third toe of his right foot without any bone destruction (Fig. 2). Ultrasonogram (Fig. 3) showed 2 partly fused hypoechoic masses measuring 1.3×1.3 cm and 1.2×1.6 cm, respectively, within the lesion. Ultrasonogram also showed that the lesion was well vascularized and contained sonolucent fluid, which was not found in the resected solid masses. Because of the slow growth of mass, we diagnosed this lesion as a benign tumor and we recommended a biopsy to confirm our diagnosis before treatment. Unfortunately, the patient declined the biopsy to confirm our diagnosis before we

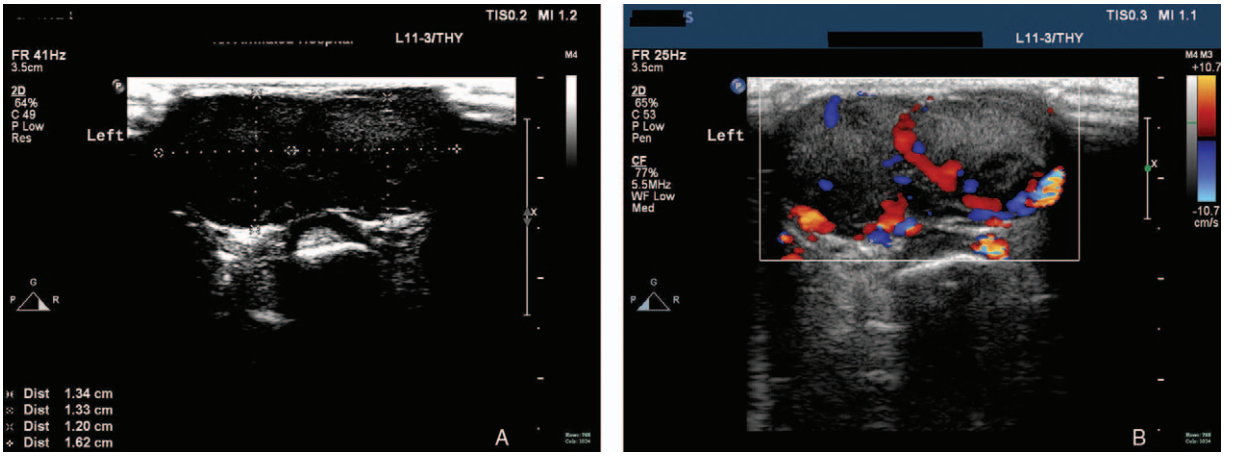

Figure 3. Preoperative ultrasonogram of the lesion in the toe. (A) The 2 partly fused hypoechoic masses measuring 1.3×1.3 cm and 1.2×1.6 cm. (B) Sonolucent fluid was seen in the lesion, and the lesion was well vascularized.

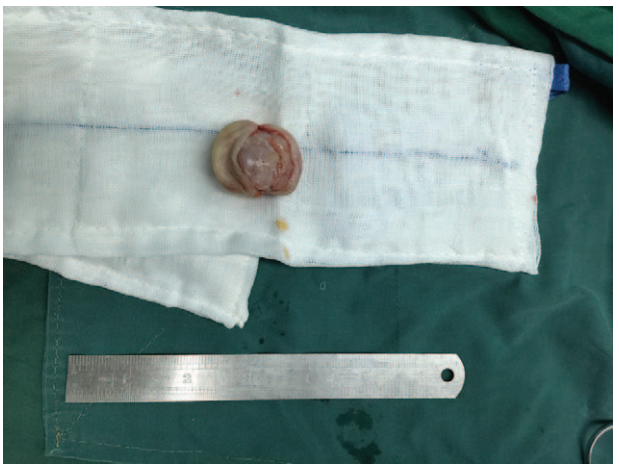

Figure 4. Intraoperative photograph shows that the lesion is composed of 2 masses, and together was 2.3 × 1.5 × 1.2 cm. The lesion has no capsule.

amputated the toe with the tumor under local anesthesia with the patient's approval. During the operation, we found that the lesion occupied almost the entire space of the toe tip (Fig. 4) and the lesion was a bilobed mass measuring 2.3 × 1.5 × 1.2 cm without a capsule. Histological findings confirmed that the tumor was benign CS with atypical neoplastic cells that had large hyperchromatic and pleomorphic nuclei (Fig. 5). Immunohistochemical (IHC) staining showed that tumor cells were S-100 positive (Fig. 6A), pan-cytokeratin positive (Fig. 6B), melan-A negative, CD34 positive, smooth muscle actin (SMA) negative, desmin negative, mostly Ki-67 negative (<5% cells positive), P63 negative, EMA (epithelial membrane antigen) mostly negative (a few positive cells), CD117 negative, G15 (gross cystic disease fluid protein 15) mostly negative (a few positive cells), and MMG (mammaglobin) negative (data not shown). The surgical incision wound healed well without any infection. Two years after the surgery, the patient had not experienced any tumor recurrence.

Ethical approval for this report was granted by the Medical Ethics Committee of the First Affiliated Hospital College of Medicine, Zhejiang University.

3. Discussion

CS, also known as a mixed tumor of the skin, is a rare, benign tumor of skin, usually occurring in the head and neck region.[1,3] Cells of sweat gland and ectopic salivary gland are the origin of a CS tumor.[3,4] CS was first described by Hirsch and Helwig in 1961 when they found the sweat gland elements within a cartilaginous stroma of CS.[5] Less than 0.1% of all skin tumors are diagnosed as CS.[1] Malignant CS occurs mostly in extremities, as does benign CS, but is much more rare.[2] Unlike benign CS, malignant CS grows quickly and invasively, and recurs frequently after resection.[6–8] Some factors that predict malignancy of CS tumor are known. A CS tumor size >3 cm is associated with increased risk of malignancy.[9] Excessive mucoid

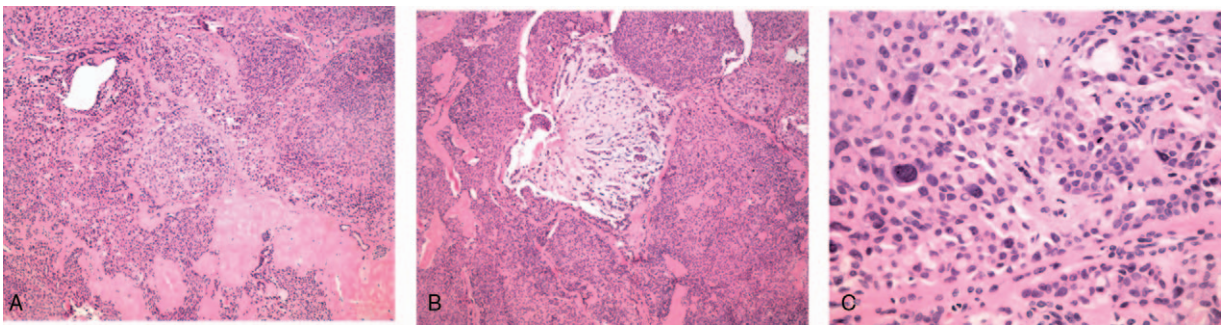

Figure 5. Pathological examination of tumor using hematoxylin–eosin staining. A. Tumor cells are arranged in nodular structures with glass-like changes and glandular tube structures in the intercellular matrix (original magnification 100×). (B) Focal myxoid in the chondroid intercellular matrix (original magnification 100×). (C) Tumor cells vary in size and exhibit significant nuclear atypia and frequent mitoses (original magnification 400×).

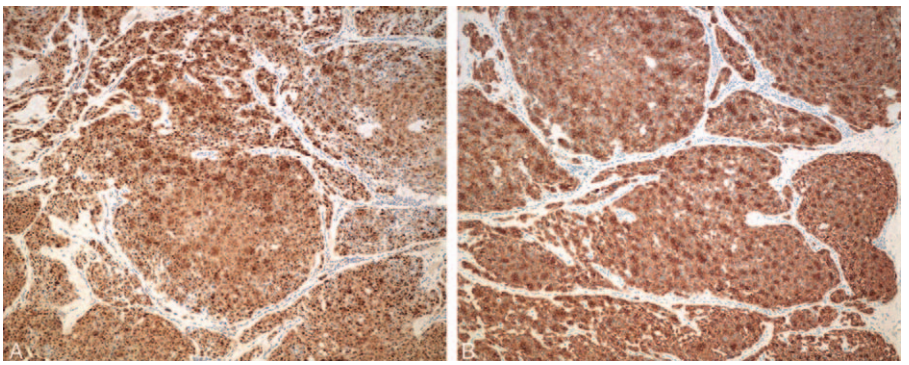

Figure 6. Immunohistochemical analysis of tumor showing that tumor cells are S-100-positive cells (A) and pan-cytokeratin-positive cells (B) (original magnification 100×).

matrix, numerous mitoses, and poorly differentiated chondroid components are important indicators of malignancy.[2,10] The histological features criteria of malignant CS are cytologic atypia, infiltrative margins, satellite tumor nodules, tumor necrosis, and involvement in deeper tissues.[11,12] The CS tumor in our case was located in the toe tip and the greatest dimension of the resected tumor was nearly 3 cm, therefore, we must suspect the possibility of its malignancy. In addition, histological examination showed that the cells in the tumor were atypical neoplastic cells with large hyperchromatic and pleomorphic nuclei, suggesting active growth. Furthermore, IHC staining showed that cells were S-100 positive, pan-cytokeratin positive, melan-A negative, CD34 positive, SMA negative, desmin negative, Ki-67 mostly negative (<5% cells positive), P63 negative, EMA mostly negative (a few positive cells), CD117 negative, G15 mostly negative (a few positive cells), and MMG negative, suggesting that the mass originated from myoepithelial cells.

Cutaneous myoepithelioma is a rare benign tumor with prominent myoepithelial cells, yet shares histopathological features with CS.[13] Cutaneous myoepithelioma of the salivary glands is the most common known. No ductal or syringomatous epithelial structures are observed compared with CS (lacking ductal differentiation).[14,15] By immunohistochemistry, cutaneous myoepithelioma was reactive for epithelial markers (keratins, epithelial membrane antigen).[16] Malignant myoepithelioma, also as myoepithelial carcinoma, is a rare salivary gland tumor composed of myoepithelial differentiation cells.[17] It also can be found in the bone,[18] soft tissue,[19] nasopharynx,[20] lung,[21] and bronchus.[22] The characteristic of malignant myoepithelioma was locally aggressive, nerve involvement, occasional regional lymph node involvement, and eventually metastasized.[23] Proliferating myoepithelial cells, mitoses, and tumor necrosis were also observed in microscopic findings.[24] EWSR1-ZNF444 rearrangement,[25] C-kit,[26] nuclear accumulation of p53 and cyclin D1,[27] and platelet-derived growth factor A genes[28] are the defining pathogenetic feature of malignant myoepitheliomas. Other differential diagnoses of all other cutaneous tumors should be included, such as neurofibromas, epidermoid cysts, mucinous cysts, and lipomas.[1] The diagnosis of CS is ultimately confirmed by histopathological examinations, but a needle aspiration biopsy is also helpful for diagnosis before the surgery. The diagnosis, benign CS, in our case, was confirmed by HE staining and IHC staining.

Treatment options of CS include resection, chemotherapy, and radiotherapy. Complete excision of the tumor is the recommended treatment due to the malignant potential of the tumor. In addition, one may fail to totally resect the lesion in cases like ours of multiple CSs in 1 digit, so preoperative imaging that can help identify lesions of multiple CSs needs to be performed, thereby guiding resection. For example, radiography failed to show the 2 partially fused masses of the tumor that were later shown by ultrasonography in our case. Adjuvant chemotherapy and radiotherapy are effective to decrease recurrence after resection.[6,29,30] Local radiotherapy is effective to prevent skeletal metastasis.[2] However, our case was not treated either with adjuvant chemotherapy or radiotherapy. Our case did not have any recurrence during the 2-year period of follow-up.

4. Conclusion

CS growing in extremities and having large tumor size are associated with higher likelihood of malignancy. Here, we show that CS can also present as a bipartite mass in a toe with its greatest dimension close to 3 cm. Complete surgical resection and long-term follow-up are essential in the management of these cases.

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