J Hear Sci, 2020; 10(2): 67–70
DOI: 10.17430/JHS.2020.10.2.8

A RARE CASE OF CONGENITAL SUPRASTERNAL DERMOID CYST

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Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

Abstract

Background: Dermoid cysts are benign congenital lesions occurring in about 80% in the ovaries and testicles and only in about 1.6-7% are they found in the head and neck region, mainly in the midline of the body.

Case report: A rare case of congenital dermoid cyst in the central compartment of the neck is presented. It was diagnosed when the patient was 10 years old; later at age 22 the patient was re-diagnosed and qualified for surgical treatment. The paper outlines the diagnostic and therapeutic procedures performed. It demonstrates the value of appropriate tests and exclusion of other possible diseases.

Conclusions: Dermoid cysts are a rare disorder affecting mainly the ovaries and testicles which require detailed diagnostics as a part of differential diagnosis. In most cases, they do not cause severe symptoms and often require only simple surgical treatment.

Key words: diagnostics • treatment • congenital dermoid cyst

RZADKI PRZYPADEK WRODZONEJ NADMOSTKOWEJ TORBIELI SKÓRZASTEJ

Streszczenie

Wprowadzenie: Torbiele skórzaste są łagodnymi wrodzonymi zmianami występującymi w około 80% przypadków w jajnikach i jądrach, a tylko w około 1,6-7% występują w okolicy głowy i szyi, głównie w linii pośrodkowej ciała.

Opis przypadku: Przedstawiono rzadki przypadek wrodzonej torbieli skórzastej w środkowym przedziale szyi. Pacjent został zdiagnozowany, gdy miał 10 lat, a następnie w wieku 22 lat został zakwalifikowany do leczenia chirurgicznego. W pracy przedstawiono przeprowadzone procedury diagnostyczne i terapeutyczne, z uwzględnieniem wartości poszczególnych testów i diagnostyki różnicowej.

Wnioski: Torbiele skórzaste są rzadkim zaburzeniem, które dotyczy głównie jajników i jąder. Wymagają szczegółowej diagnostyki różnicowej. W większości przypadków nie powodują ciężkich objawów i często wymagają jedynie prostego leczenia chirurgicznego.

Słowa kluczowe: wrodzona torbiel skórzasta • diagnostyka • leczenie

Introduction

Congenital neck defects are most often cysts. Congenital neck cysts include: thyroglossal duct cysts, branchial cleft cysts, and dermoid cysts. Hem- or lymphangiomas are less common and usually manifest as soft tumors on the neck [1]. Thyroglossal cyst is the most common congenital neck mass and occurs in 7% of the population [2]. They occur due to failure of thyroglossal duct to involute and atrophied thyroglossal duct cysts often occur in pediatric patients. The majority of them are found in the infrahyoid region [3].

The other type of cystic lesion are branchial cleft cysts. Five pairs of branchial clefts are present in a 6-week-old human embryo from which develop the organs of the head, neck, and chest. Branchial cleft cyst is a remnant of embryonic development resulting from a failure of obliteration of one of the branchial clefts. The exact location of the cyst depends on the branchial cleft from which the cyst originates. First branchial cleft cysts originate in the preauricular region or in the angle of the mandible and extend to the external auditory canal. Second branchial cleft cysts are found along the anterior border of the sternocleidomastoid muscle and sometimes drain into the patent duct to the throat. Third and fourth branchial cleft cysts are located above the sternum or collarbone. Remnants of an ultimobranchial body from the fourth and fifth branchial cleft cysts (according to some authors the fifth pair does not exist; it is rudimentary or is part of the fourth pair) are found in the thyroid in the form of fragments of cartilage or thymus tissue [4].

Dermoid cysts are benign congenital lesions occurring in about 80% in the ovaries and testicles and only 1.6–7% are found in the head and neck region, mainly in the midline of the body [5,6]. Most of these lesions found in the cervicofacial region are congenital and they are derived from entrapment of epithelial cells during midline fusion in embryonic development [7,8].

The aim of this study was to present a rare case of congenital suprasternal dermoid cyst.

Case report

A male aged 10 reported to the outpatient department for an ENT appointment due to a palpable tumor in the supraclavicular notch. The child did not report any complaints except itching during palpation. Furthermore, a laryngological examination revealed no deviation from the normal condition.

An ultrasound of the neck showed a symmetrical thyroid gland, correctly located in the neck, homogeneous parenchyma, and normal echogenicity. Focal lesions were not observed. Right lobe 36 × 9 × 10 mm; left lobe 37 × 11 × 11 mm. The isthmus was narrow. A round, firm, homogeneous, well-circumscribed (probably encapsulated) 16 mm focal lesion was revealed in the suprasternal region just under the surface of the skin, not connected with adjacent structures.

Surgical excision of the lesion was not applied due to the possibility of damage to adjacent structures which in turn could disturb the normal development of the child. Further observation of the lesion was recommended. The patient made the decision to remove the cyst after reaching maturity, 12 years after the first diagnosis.

The patient, aged 22 reported to the General Surgery Department. He did not report any complaints, except for a feeling of pressure at the lesion, which escalated with increase in the intensity of physical effort. Ultrasound of the neck (Figure 1) was performed and it revealed normal thyroid and homogeneous parenchyma; just under the skin in the supraclavicular notch, a visible, encapsulated, well-circumscribed lesion with high echogenicity of size 53 × 30 × 17 mm was seen, most likely an atheroma. No other lesions or enlarged lymph nodes were visible in the surrounding soft tissues.

Fine-needle aspiration biopsy of the lesion was performed and cytology revealed benign layers of epithelial cells and keratin. After the biopsy, the patient was referred to the Maxillofacial Surgery Department, where a contrast-enhanced CT scan was performed to highlight the lesion better and to limit possible complications resulting from the possibility of damage to neighboring structures, including blood vessels (Figure 2).

After the examinations, the patient was qualified for surgery under general anesthesia with endotracheal intubation and perioperative antibiotic treatment. Surgical treatment was performed in the Department of Maxillofacial Surgery. Description of the procedure: under general endotracheal anesthesia, with intubation through the mouth, a midline neck incision was made from the level of the thyroid cartilage to the cervical notch. After detachment of the infrahyoid muscles, the 35 × 30 × 17 mm cyst was reached and completely removed. The cyst did not adhere to the thyroid gland.

Surgery was successful, and the postoperative course uneventful. Histopathological examination of the removed

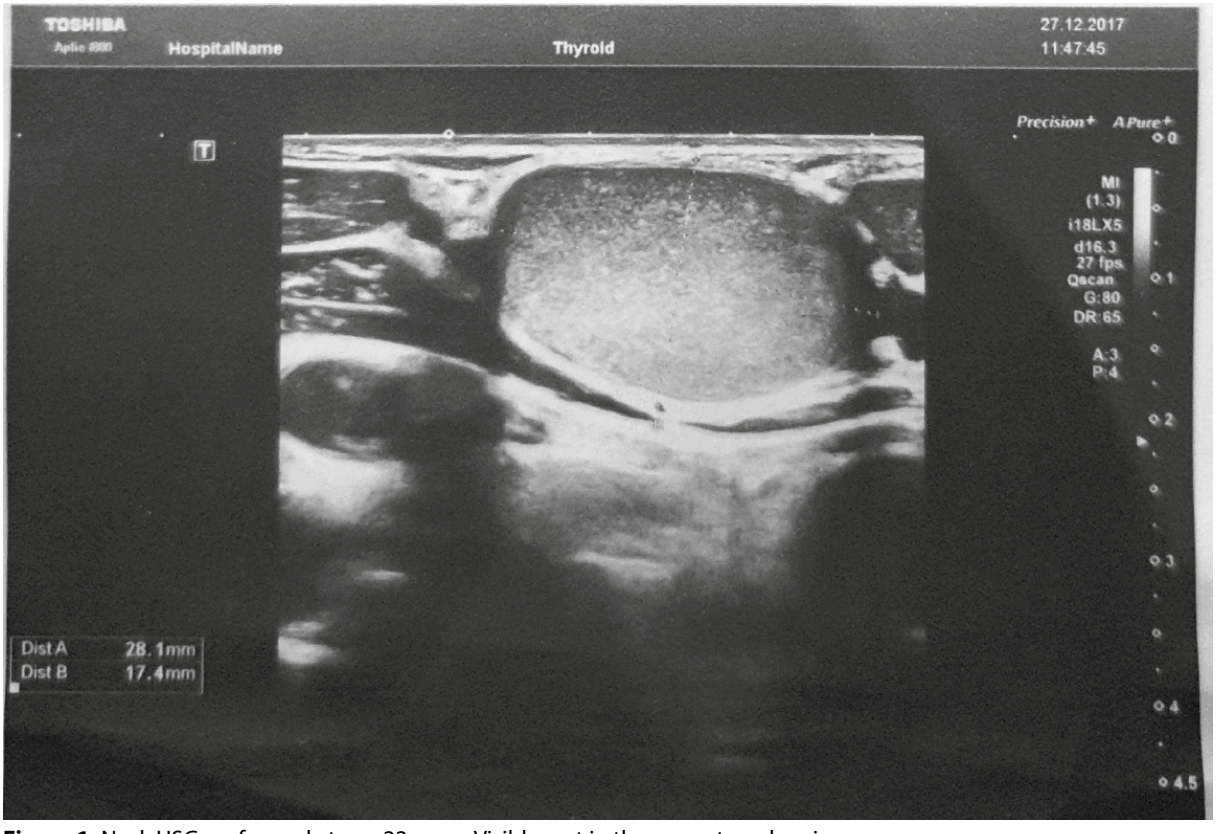

Figure 1. Neck USG performed at age 22 years. Visible cyst in the suprasternal regions

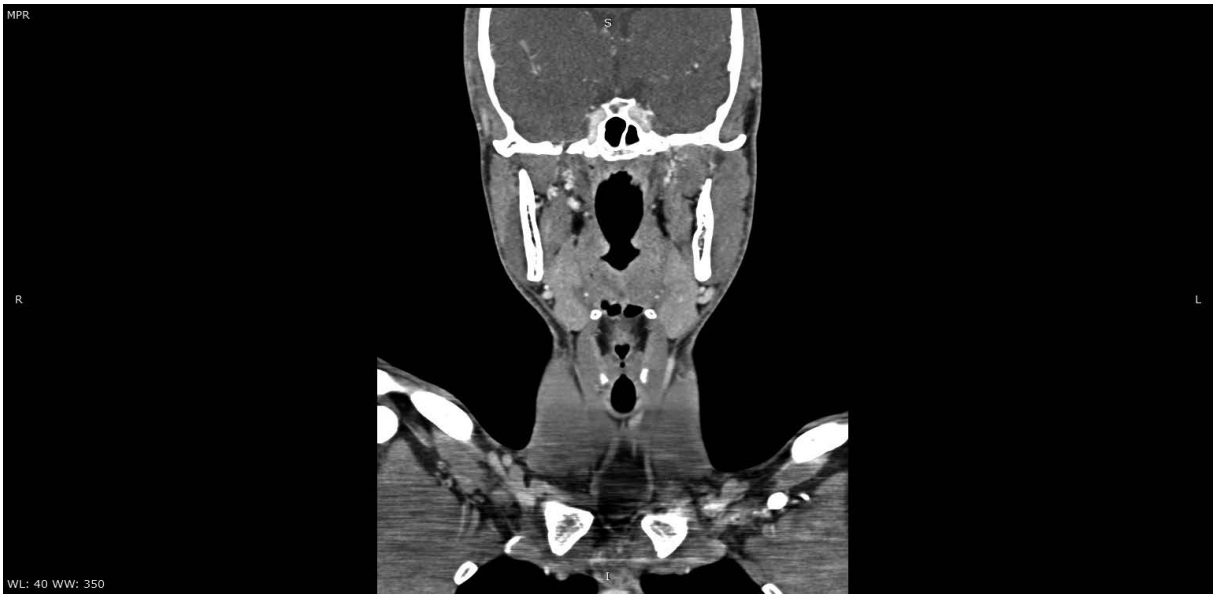

Figure 2. Computed tomography of facial skeleton and neck (frontal projection) of the patient at age 22 years with visible dermoid cyst in suprasternal regions

lesion confirmed that it was a congenital dermoid cyst of the neck. Histopathological diagnosis is shown in Figure 3. It shows a dermoid cyst with external focal adipose tissue necrosis; there is a multicellular granulomatous reaction (after BAC) at the same level inside the cyst. Surgical incision lines remained free of the pathological lineage. The cyst was filled with porridge-like masses.

Discussion

Common causes of midline neck masses include thyroglossal duct cysts, lymphadenopathy, dermoid cysts, and various odontogenic anomalies. Thyroglossal duct cysts (TDCs) often occur in pediatric patients;however, at least half are found in the second decade of life and they can also present later in adulthood [8]. The classic presentation of TDC is a midline, non-tender, palpable mass that moves with swallowing and elevates on protrusion of the tongue [9]. Thyroglossal duct cysts are usually 2–4 cm in diameter and gradually increase in size [10]. They may enlarge rapidly after an upper respiratory tract infection.

A dermoid cyst is an encapsulated cyst covered by a thin layer of keratinized cells containing adnexal structures of the skin, such as sebaceous gland, sweat gland, hair follicles, and hair [11]. Its location, size, and clinical presentation are extremely variable, thus causing confusion associated with medical conditions and emergencies requiring urgent surgical intervention due to ruptures and sprains, intestinal and bladder injuries, pregnancy, etc. [12]. The most frequently reported locations for the occurrence of a dermoid cyst within the neck and face are periorbital, nasal, submental, and suprasternal regions [13,14].

Medical history related to the lesion as well as physical examination of the head and neck are necessary to make a diagnosis. The case record should include the onset of the lesion, its duration, changes in appearance or symptoms, the presence of more lesions, associated pain, dysphagia, and fluid leakage. The patient should be examined for the presence of other syndromes. A detailed physical examination can be helpful in excluding possible disease entities. Moreover, imaging plays an important role in determining the nature and extent of tumor tissue. A methodical approach to imaging of tumors should include determining the sites of their primary origin, which are related to the anatomical spaces of the head or neck and contain characteristic imaging features. Ultrasound and MRI are the imaging methods of choice in the pediatric population. Computed tomography is another valuable method that can also be included but its inherent ionizing radiation leads to more limited use in children. Each of these three imaging methods has advantages and disadvantages. The management of dermoid cysts depends on their location. Most cysts only require a simple excision, although sometimes central dermoid cysts can move deeper into the tissue and require more extensive surgery [15].

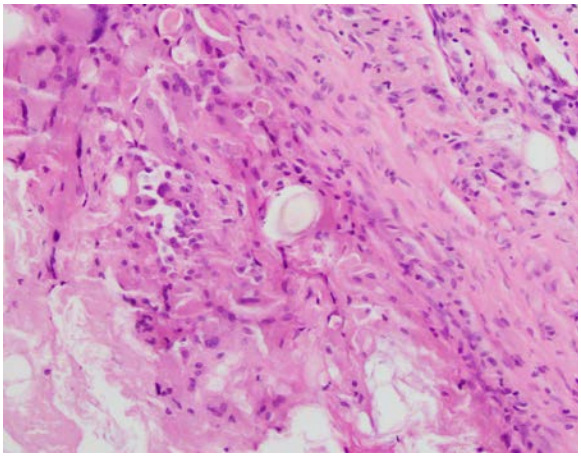

Figure 3. Microscopic image of dermoid cysts with granulation visible within its central part and a hair shaft (10x magnification)

Conclusions

Dermoid cysts of the neck are a rare disease affecting mainly the ovaries and testicles. They require detailed diagnostics to exclude many disease entities. In most cases, they do not cause severe symptoms and often require only simple surgical treatment.

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