

C A S E R E P O R T

Dermoid Cyst of the retroauricular region: a rare clinicopathological entity

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Abstract. *Background and aim:* Dermoid cysts (DC) of the head and neck are rare congenital anomalies derived from entrapment of ectodermal cells at lines of fusion in the embryo into mesoderm. *Methods:* We describe a 22-years-old female with an unusual presentation of DC in the subcutaneous tissue of the retroauricular region. *Results:* The pathological examination of the surgically removed specimen confirmed the diagnosis of Dermoid Cysts. *Conclusions:* A DC in this region is rare and may be misdiagnosed as a retroauricular lymph node. Complete excision of the lesion must be achieved with pathology study to confirm diagnosis. (www.actabiomedica.it)

Key words: dermoid cyst; retro-auricular region; congenital anomaly; head and neck

Introduction

A dermoid cyst (DC) is a rare congenital lesion that generally became evident short after birth and are frequently located close to the midline (1). Histologically a keratinizing squamous epithelium is typically present together with a variable number of dermal derivatives such as hair follicles, smooth muscle, sweat and sebaceous glands, and fibroadipose tissue (2).

Clinically DC appears as a rounded and slowly growing lesion, with no specific symptoms (3). Approximately 7% of DC occur in the head and neck, with the most reported locations being periorbital, nasal, submental, lip, palate and suprasternal (2).

We report a case of a DC in the subcutaneous tissue of the retroauricular region which was misdiagnosed as a lipoma. The location is unusual with less than five cases described in English literature (1,3).

Case presentation

A 22-years-old female presented with a gradually enlarging mass in the right retro-auricular area; the mass was present since early childhood.

The patient denied any additional symptoms or traumatic and inflammatory events.

At physical examination, a 30×15 mm soft mass in right retroauricular area was evident, mobile with respect to the overlying skin. No other anomalies were found in head and neck region; audiological examination was also normal.

A radiological examination with MRI confirmed that the mass was confined to the subcutaneous tissues.

Complete surgical excision was achieved under local anesthesia. After raising the skin flap, a pinkish encapsulated mass was observed, in contact with the bony surface of the temporal bone.

On pathological examination the mass measured 30×15 mm, the walls were lined with keratinized squa-

mous epithelium containing hair follicles and sebaceous glands; the definitive diagnosis was dermoid cyst (Figures 1, 2).

Twenty-two months after the operation there were no signs of recurrence.

Discussion

The pinna develops from the six hillocks of His, which form from the first and second branchial arches. Defective closure of the first branchial cleft or failure

of fusion of the primitive ear hillocks may result in the formation of small pits, sinuses or fistulae in front of the pinna or cystic lesions occurring in the external ear as a dermoid cyst, epidermoid cyst, cystic teratoma, branchial cyst and trichilemmal cyst (1;4).

Dermoid cysts (DC) are lesions composed of two germ layers, i.e. ectoderm and mesoderm. Both dermoid and epidermoid cysts are ectoderm-lined inclusion cysts; however, while epidermoid cysts have only squamous epithelium, dermoid cysts also contain hair, sebaceous and sweat glands (5).

In contrast to teratoid cyst, dermoid cyst lacks endodermal derivatives (e.g., gastrointestinal, or respiratory mucosa and smooth muscle) (6).

Pryor et al performed a retrospective review of the presentation, diagnosis, treatment, and outcomes of pediatric dermoid cysts of the head and neck examined between 1980 and 2002 in a single institution (2); forty-nine patients (59% girls) had a DC of the head and neck (median age at diagnosis was 22 months), with only two cases in the retro-auricular region (2%). Horikiri described a 6-year-old female with a DC in the right postauricular area (3).

In literature, there are also few reports of malignant transformation, mainly in ovaries and testes (7). Squamous cell carcinoma is the most common malignancy diagnosed in dermoid cysts followed by adenocarcinoma (8). In these cases the treatment of choice is an extensive surgery with negative surgical margins and combination of chemo-radio therapy; despite the treatment, many such cases present a poor prognosis (7, 9). Prolonged chronic inflammation is indicated as a factor for malignant transformation.

DC may occur anywhere in the body. They primarily occur in the gonads; however, they also occur at extragonadal sites along the midline of the body. The head and neck region is a rare location for such lesions in children or adults. The most common sites include midline neck or nose, nasolabial fold, and lateral third of the eyebrow (along the embryological closure lines) (2). In the temporal bone area DC have been described in different sites, including the middle ear (10), mastoid process (1), Eustachian tube (11) and petrous apex (12).

Dermoid cysts generally present with slow and progressive growth. On MRI imaging, DC have variable characteristics on T2 sequences, depending on the

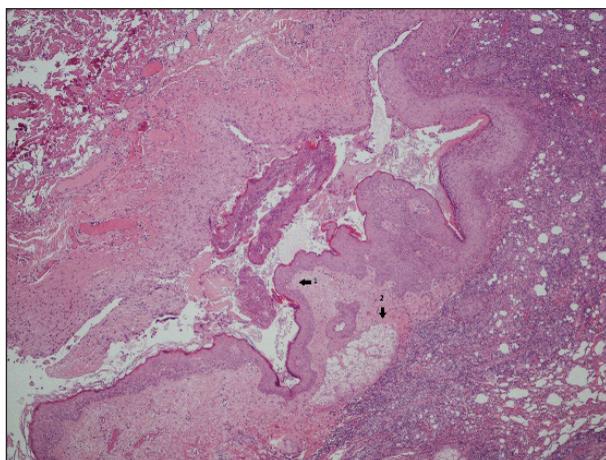


Figure 1. Dermoid cyst lined by keratinized stratified squamous epithelium (arrow no.1) and pilosebaceous units (arrow no. 2) embedded in the cyst wall (H&E; x100). H&E, hematoxylin and eosin.

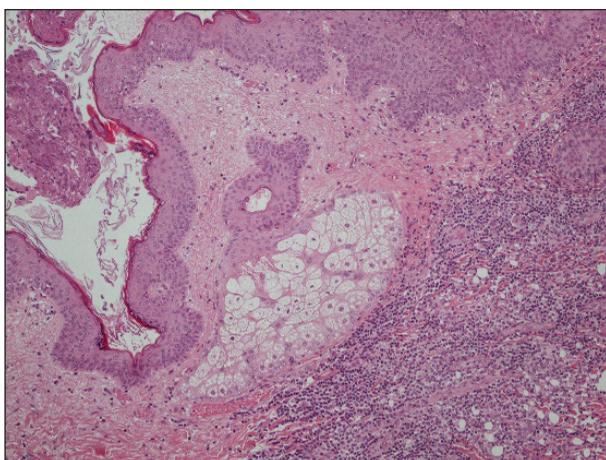


Figure 2. Higher magnification of pilosebaceous units. The interstitial tissue contains squamous epithelium, muscle, hair roots, sebaceous glands and sweat glands.

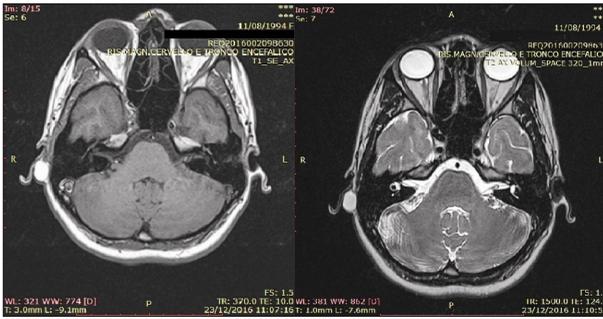


Figure 3. MRI: a) T1 image (hyperintense, due to cholesterol components); b) T2 image (less hyperintense).

water content; on T1 they are more frequently hyperintense (due to cholesterol components). Hyperintense droplets may be visible on T2 if rupture has occurred. They usually do not enhance after gadolinium infusion and on DWI sequences, DC is less likely to show diffusion restriction than epidermoid cyst (13). On CT imaging, typically DC appear as well-defined fat density lobulated masses. Calcifications may be present in the wall. Enhancement is uncommon, and if present should be a thin peripheral rim (14) (Figure 3).

The most common reason for surgical removal of dermoid cyst of the peri-auricular area is cosmetic. Like dermoid cysts in other areas the goal of the surgery is the complete excision in order to reduce the risk of recurrence (15).

Conclusion

DC in the retroauricular area is a rare benign lesion, often present since childhood. Recurrence is unusual after complete excision. Malignant transformation, although rare, mandates for curative treatment of this type of lesion at an early stage.

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