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CASE REPORT

Fine Needle Aspiration Cytology diagnosis of the Eccrine Variant of Chondroid Syringoma- Case Report of a Rare Entity with Review of Literature

BHASIN T S * , MANNAN R ***, BHATIA P K ****, SHARMA M ***, BHALLA A *****

ABSTRACT

Chondroid syringomas (CS) are rare mixed tumours of sweat-gland origin which were first described by Billroth in 1859, that have both benign and malignant forms. They are also known as mixed tumours of the skin and are composed of both epithelial and mesenchymal components. A 40-year-old female presented to the surgical outpatient department with a gradually increasing swelling over the side of the nose, of 1 year duration. FNAC was done and the aspirate was thick and mucoid. Microscopic examination revealed clusters of epithelial and myoepithelial cells, embedded in a metachromatic, chondromyxoid ground substance. Based on these features, a diagnosis of chondroid syringoma was made and histopathological examination was advised. On histopathological examination, it was found that there were numerous nests of polygonal cells and interconnecting tubuloalveolar structures whicha were lined by a single layer of cuboidal epithelial cells. The stroma consisted of cells in a bluish chondroid matrix. Based on these findings, a diagnosis of the eccrine variant of CS was given. The patient was well after excision and no recurrence was reported. CS is a rare primary skin tumour; the incidence is < 0.098% and affects middle aged and older men. The most common sites are the head and neck region, the hand, foot, the axillary region, the abdomen, penis, vulva and the scrotum. Fine needle aspiration cytology has been used for diagnostic purposes and may prove to be useful to determine the pathology before excision of the tumour. However, examination of excised tissue is most reliable in establishing a definitive diagnosis.

Key Words: Chondroid syringoma, eccrine, FNAC.

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Introduction

A chondroid syringomas (CS) are rare mixed tumours of sweat-gland origin which were first described by Billroth in 1859, that have both a benign and malignant form. They are also known as mixed tumours of the skin and are composed of both epithelial and mesenchymal components. Hirsch and Helwig gave them the appellation *chondroid syringoma,* because of the presence of sweat gland elements which are set in a cartilaginous stroma [1].

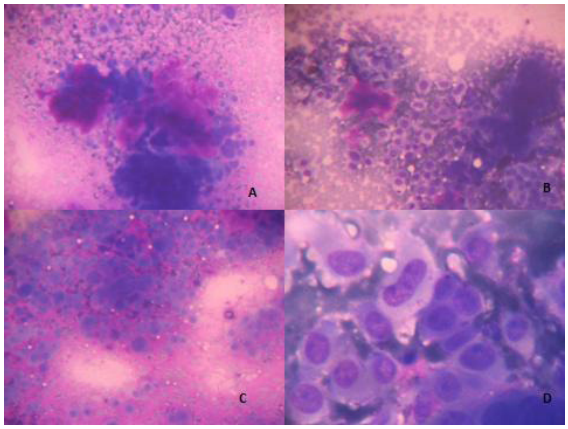
The incidence of CS is low and has been reported at 0.01-0.098 percent. The lesions are typically located on the head and neck and are non-ulcerating, slow-growing, subcutaneous, or dermal nodules [1], [2], [3]. We present here, a rare case of the eccrine variant of chondroid syringoma, occurring on the side of the nose in a 40 year old woman. There are only a few case reports describing the fine needle aspiration

cytological (FNAC) features of chondroid syringoma and that too, the eccrine variant, which itself is very rare. The diagnosis was established by FNAC and confirmed by histopathology.

Case Report

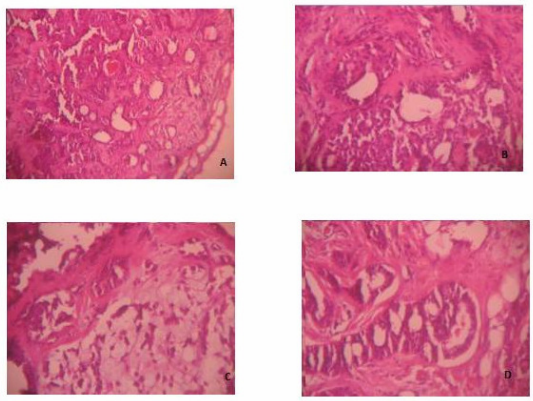
A 40-year-old female presented to the surgical outpatient department with a gradually increasing swelling over the side of the nose, of 1 year duration. Physical examination revealed a mobile, firm swelling that measured 0.8 cm in diameter. FNAC was done with the help of 22 G needles. Two to three passes at two different sites were done. The smears prepared, were stained with May-Grunwald-Giemsa (MGG) and Papanicolaou stains. Grossly, the aspirate was thick and mucoid. Microscopic examination revealed clusters of epithelial and myoepithelial cells embedded in a metachromatic, chondromyxoid ground substance [Table/Fig 1]. The epithelial cells had round, monomorphic nuclei having dispersed chromatin, with a moderate to abundant amount of cytoplasm. Based on these features, a diagnosis of chondroid syringoma was made and histopathological examination was advised.

At the time of excision, gross pathological analysis revealed well-encapsulated tan coloured tissue. No areas of haemorrhage or necrosis were noted. The histopathological examination revealed a well-circumscribed proliferation of epithelial cells, interspersed with foci of myxoid stroma. There were numerous nests of polygonal cells and interconnecting tubuloalveolar structures lined by a single layer of cuboidal epithelial cells [Table/Fig 2]. The stroma consisted of cells in a bluish chondroid matrix. Based on these findings, a diagnosis of the eccrine variant of CS was given. The patient was well after excision and no recurrence was reported.



(Table/Fig 1)

Fig 1 A and B- Clusters of epithelial and myoepithelial cells embedded in a metachromatic, chondromyxoid ground substance (200 X Giemsa).
Fig 1 C- The clusters of epithelial cells and myoepithelial cells (200 X Giemsa).
Fig 1 D- Same as 1 C with higher magnification (400 X Giemsa).



(Table/Fig 2)

Fig 2 A- Well-circumscribed proliferation of epithelial cells (100 X H&E).
Fig 2 B- Same as 2A with higher magnification (200 X H&E).
Fig 2 C- Foci of myxoid stroma (400 X H&E).
Fig 2 D- Interconnecting tubuloalveolar structures lined by single layer of cuboidal epithelial Cells (400 X H&E).

Discussion

CS is a rare primary skin tumour; the incidence is < 0.098% and affects middle aged and older men. [4],[5]. The most common sites are the head and neck region, hand, foot, the axillary region, the abdomen, penis, vulva and the

scrotum. CSs are a group of tumours of the salivary gland that contain varying amounts of mucoid and cartilaginous material [1]. The first case of chondroid syringoma is believed to have been reported by Nasse in 1892 [6]. Virchow and Minssen [2] referred to them as mixed tumours with both epithelial and mesenchymal origins. Histologically, CS consists of mixed epithelial and mesenchymal elements, with epithelial cells arranged in cords and forming tubules with a myoepithelial layer, set in myxoid or chondroid stroma [2],[7], [8], [9]. Two histological variants of this tumour are described, the eccrine type with smaller lumens, lined by a single row of cuboidal epithelial cells and the apocrine variant with tubular and cystic branching lumina, lined by two rows of epithelial cells [2].

Headington recognized 2 types, apocrine and eccrine. The apocrine type demonstrates irregular branching tubules (tubulocystic pattern) lined by at least 2-cell-thick epithelium. The eccrine type is characterized by rather uniform, small, round tubules that are evenly spaced within a myxoid-chondroid matrix. 10 CS lesions usually were not clinically distinctive and the diagnosis was made on the basis of microscopic examination [2],[6]. Also, there is limited literature on the degree of the aggressive nature and the propensity of the recurrence of the eccrine variant over the apocrine variant of CS.

The tubuloalveolar components of chondroid syringomas are composed of two layers of cells with different immunophenotypes. The inner layer expresses epithelial markers such as cytokeratin [11]. The outer layers express mesenchymal markers such as vimentin, S-100 protein, neuron-specific enolase (NSE) and glial fibrillary acidic protein (GFAP) [11]. Initially, it was thought that the chondroid area of the tumour showed ultrastructural features which were identical to myoepithelial cells, a component of the outer layer of cells [12]. However, subsequent studies have concluded that the cartilaginous matrix is true cartilage produced by ultrastructurally typical chondrocytes and not by the pseudocartilage which is produced by the myoepithelial cells [11].

Clinically, the differential diagnosis of CS is made with other benign tumours of the epidermal or mesenchymatous appendages such as the dermoid or sebaceous cysts, neurofibroma, dermatofibroma, basal cell carcinoma, pilomatricoma, histiocytoma and seborrheic keratosis [4]. Therefore, CS should be considered in the differential diagnosis of any sub cutaneous nodule in the head and neck region of a middle aged individual, especially in males.

Because of the risk of malignancy, the first-line treatment is total excision, where a cuff of normal tissue is necessary to avoid recurrence [2]. This is followed by regular follow-up, to look for local recurrence and any feature of malignancy [13]. The recurrent lesion can be treated by surgical re-excision [14] .Malignancy in CS is rare, with reported cases occurring in young female patients in the extremities and the torso [15] .

Fine needle aspiration cytology has been used for diagnostic purposes and may prove to be useful to determine the pathology before excision; however, examination of the excised tissue is most reliable in establishing a definitive diagnosis [16],[17].

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Key Message

1. Chondroid syringoma is rare primary skin tumour; the incidence is < 0.098% and affects middle aged and older men. Most common sites are the head and neck region, hand, foot, the axillary region, the abdomen, penis, vulva and the scrotum.
2. The aspirate on FNAC is usually thick and has mucoid like consistency. Smears reveal clusters of epithelial and myoepithelial cells embedded in a metachromatic, chondromyxoid ground substance.
3. On histology; 2 types of chondroid syringoma are recognized: apocrine and eccrine. The apocrine type demonstrates irregular branching tubules (tubulocystic pattern) lined by at least 2-cell-thick epithelium. The eccrine type is characterized by rather uniform, small, round tubules that are evenly spaced within a myxoid-chondroid matrix.
4. Immunohistochemically, the inner layer was found to express epithelial markers such as cytokeratin, whereas; the outer layers express mesenchymal markers such as vimentin.
5. Clinically, the differential diagnosis of Chondroid syringoma was made, with other benign tumours of epidermal or mesenchymatous appendages such as the dermoid or sebaceous cysts, neurofibroma, dermatofibroma, basal cell carcinoma, pilomatricoma, histiocytoma and seborrheic keratosis .
6. As there is a risk of malignancy in chondroid syringoma, the first-line treatment is total excision, where a cuff of normal tissue is necessary to avoid recurrence.