Branchial cysts, sinuses and fistulae

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Abstract Objective: The main aim of this paper is to study the details of pathology, diagnosis, timing of operation and outcome treatment of branchial cysts, sinuses and fistulae.

Design: It is a retrospective analysis of 12 consecutive cases of branchial cleft anomalies treated at the King Fahad University Hospital, Al-Khobar between 1983 and 1994 inclusive.

Result: The lesions were predominantly among females; their ages ranging between 5 months and 28 years. Three unilateral cysts were operated upon much earlier than fistulae and sinuses because of a degree of uncertainty as to the precise nature of the lesions. Another three cases presented bilaterally; two cases of bilateral fistulae with connection to the pharynx and a case of a fistula on one and a sinus on the other side.

The surgical excision was successful in all patients except one, who developed postoperative infection and recurrence requiring a second and more complete fistulectomy three months later.

Conclusion: The presence of squamous metaplasia in one fistular tract lined by ciliated columnar epithelium points towards the possibility of premalignant change emphasizing the necessity of early operation. The diagnosis and the management of these lesions have to be considered in the light of their embryological evolution.


Keywords: Branchial cysts, sinus, fistula. branchiogenic cysts, sinus, fistula

The branchial apparatus consists mainly of four branchial arches located on each side of the future head and neck region of an embryo. Externally the four arches are separated by three clefts which are internally matched by pharyngeal pouches separated by membranes. The branchial apparatus transforms to its adult derivatives, such as maxilla, mandible, hyoid bone, middle ear, tympanic membrane, larynx as well as muscle of mastication and of facial expression, so that it completely disappears by the end of the sixth embryonal week. Most of the congenital malformations encountered in the head and neck region occur from failure of completion of one or more of these transformational processes.

Embryogenesis of branchial lesions. The first cleft normally fills in except the part left as the eustachian tube and auditory canal. The second and third clefts fill in completely. If any part or all of a cleft or pouch or both remains patent, then we can expect to find tracts which extend from an external opening at the skin cephalad for varying distances. Some are very short and extend only one or two centimeters (sinus); others may be patent all the way running through the pharyngeal wall with an internal opening (fistula). At times only a central portion of a cleft or pouch remains patent; such lumen gradually fills with secretions to form the familiar branchial cyst (1-2) (Fig.1).

The purpose of this paper is to review the case records of patients seen in a teaching hospital in Saudi Arabia, to review the embryological background of these lesion and to make recommendations towards logical diagnosis and management.

Material and methods The case records of patients with branchial cleft anomalies who were treated at the King Fahad University Hospital from 1982 G to 1994 G were reviewed. The following information was extracted: sex, age at diagnosis and at operation, clinical presentation, cyst features and contents, operative details, pre-and postoperative complications, histological findings, treatment and outcome.

Results There were 12 patients (2 Saudi and 1 Sudanese male and 9 Saudi females) with 15 lesions, of which 3 were cysts containing serous yellowish fluid. Another three cases presented as bilateral lesions; two bilateral fistulae and a case of

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a fistula on one and a sinus on the other side. The remaining were sinuses. All sinuses and fistulae were diagnosed in the neonatal period except one sinus which was diagnosed at the age of three years.

Twelve lesions presented as a skin opening, at times discharging serous fluid, and 3 lesions presented as a mass. The sinuses and fistulae were located at the lower third, the cysts between upper and middle third of the anterior border of the sternocleidomastoid muscle. Pain was a presenting symptom in a case of infected fistula with abscess formation. All cysts were unilocular; one was associated with non-specific lymphadenopathy on the contralateral side. The fistulae were bilateral in 2 instances. Of these one was associated with multiple palatine cysts, one of which was lined by squamous epithelium while the others were lined by columnar epithelium.

The operation on 10 cases of sinuses and fistulae were delayed for an average of 5.2 years (range 5 months to 16 years); in the remaining two cases the parents have not yet consented to the surgical treatment. In contrast, the diagnosis of cyst was established at birth, and at the age of 17 and 28 years, in one case each. The operation was delayed for two years in the first two cases, and performed immediately in the third case.

The operation was carried out through a single incision in 12 lesions and through a “step-ladder” incision in one. Dissection was generally not difficult, and carried out with scissors directly on the tract. The lesions were removed completely except in one case of fistula, where incomplete removal resulted in an abscess formation and recurrence necessitating drainage with curettage followed by fistulectomy three months later. All specimens were evaluated histopathologically. Results are summarized in the table. A case of fistula with 16 years delay in management showed squamous metaplasia within the ciliated columnar epithelium.

The average hospital stay was 6 days (range 2 to 15 days).

**Table 1** - Histopathological features of cysts, sinuses and fistulae

<table>
<thead>
<tr>
<th>Epithelial lining and adjacent tissues</th>
<th>Cysts (3)</th>
<th>Sinuses and fistulae (10)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Squamous epithelium</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Columnar epithelium</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Squamous + columnar</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>Lymphoid tissue</td>
<td>2</td>
<td>8</td>
</tr>
<tr>
<td>Lymphoid follicle</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>Skeletal muscle</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>Smooth muscle</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

**Discussion** Dissection of the embryo has shown that it is possible to assign the clinical lesion to a particular part of the branchial apparatus. Sinuses and fistulae that open below the angle of the jaw and pass to the cartilage of the external auditory canal, are considered remnants of the first branchial cleft. Those with an external opening at the anterior border of the lower third of sternocleidomastoid and pass between the internal and external carotid arteries, are remnants of the second cleft (Fig.1). The remains of the third cleft pass lateral to both vessels, or behind the internal carotid artery to open in the pyriform sinus. The presence of thyroid or thymus tissue adjacent to the tract may support its origin from the third or even fourth branchial cleft. On the other hand, some different clinical and pathological characteristics have been highlighted in an attempt to formulate a new definition and classification. Most branchial remnants arise from the second branchial cleft; those derived from the first cleft are rare. Lesions

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**Fig. 1:** Diagram depicting most frequent locations of branchial cysts, sinuses and fistulae.
derived from the third branchial cleft are extremely rare and have recently been more frequently reported.2,8,10 The twelve cases reported here are considered derivatives of the second branchial cleft. Of these 12 patients 11 are Saudis; these 11 cases are the first ever reported. Fistulae and sinuses present as tiny openings, at times discharging serous yellowish fluid. They usually are discovered by the mother at birth or in early infancy, while most cysts are found in older children and adults.2,6,7 This trend is reflected well in this series. The only sinus that was recognized first at the age of 3 years may have been a cyst, which underwent subclinical infection and perforation resulting in sinus formation.

Air-filled lateral cervical cysts have been reported as bronchogenic11 or branchial cysts.9 In rare instances branchial fistulae present in association with defects of the external, middle or inner ear, stenosis of the tear duct, as well as renal anomalies.12,13 Presence of branchial fistulae or sinuses should therefore stimulate the search for this dominant autosomal inherited syndrome because of major consequences of management and outcome of this condition.

Interestingly there was a female preponderance (75%) in this series, although, sex difference has not been reported.5,9

Preoperative diagnosis was suggested by the presence of lateral cervical fistulae or cysts and confirmed by operative finding and histopathological examination. However, in some cases of sinuses with short tracts, it was difficult to assign the anomaly to a particular branchial cleft with certainty.

Cystic lesions were operated upon much earlier than the fistulae (on the average 1.3 vs 5.2 years) reflecting the uncertainty in the diagnosis of cystic conditions: the differential diagnosis of branchial cyst includes metastatic carcinoma, lymphoid tumors, cystic hygroma, carotid body tumor, dermoid cyst, parotid tumor, thyroid tumor and deep-seated lipomas.14

Pathologic findings in this series were similar to those reported by others.6,16 A case of a cyst lined by columnar epithelium and another of a sinus with smooth muscle found adjacent to its tract were the exception. Both cysts and sinuses can theoretically be lined by squamous or columnar epithelium or by both according to their likely origin from a branchial cleft (ectoderm) or a branchial pouch (endoderm) or from both. The presence of smooth muscle in the wall of a sinus can be explained by the existence of pluripotent cells in the branchial apparatus that can transform to lymphoid tissue, lymphoid follicle, skeletal or smooth muscle. Complete extirpation of the lesion is mandatory. It is easily achieved if undertaken at an early stage before infection sets in. The operation is indicated both for cosmetic reasons and to prevent infection which may be encountered in 25-30% of cases.14,15

The existence of primary neoplastic change in a branchial cleft anomaly is controversial. However, cases of branchial anomaly with query primary carcinoma have been reported.16 Squamous metaplastic change in a long standing fistula lined by columnar epithelium was observed in this series and reported by others.17 This suggests that the list of indications for operative removal should include the possibility of malignant change.

It is usually advocated that a step-ladder approach be used in excising these lesions.17 But I think like some2,5 that most cases in children can safely be operated on through a single incision, particularly in the absence of repeated episodes of infection and if the tract is not too long. Nevertheless careful dissection is important to protect the vital structures such as jugular vein, carotid arteries, vagus and hypoglossal nerve.17 Aspiration of the cyst can be helpful in access to deep surfaces, and prevention of rupture of the cysts.18 The facial nerve is at high risk in the excision of remnants of the first branchial cleft. In fact, a formal superficial conservative parotidectomy with exposure of all main divisions of the facial nerve has been recommended.19 The delay of surgical treatment is advisable if the lesion is diagnosed soon after birth or to control infection.

References
ملخص:

هدف هذه الورقة هو دراسة تفاصل التشريح المرضي والتشخيص، وتحديد الموعد المناسب للمداخلة الجراحية، ونتائج علاج الأكياس والجيوب والأنسجة الخشوية.


نتائج: تواجه الأعراض المرضية أكثر عند الإناث، وتراوحت أعمار المرضى ما بين 5-80 سنة، واجب العمليات الجراحية لثلاثة أكياس متواجدة في وقت أقرب لليومية بالجيوب والنواسير، وذلك بسبب عدم التأكد من تلبية الأفلا، واجب حالات بالجاليتين منهم حاليين بناوسير على الجاليين متصلا بالبلعوم وحالات ناسور على الجانب.

وجب على الجاليان الآخر.

وكان الاستلال الجراحي ناجحا في جميع الحالات، سواء أ جيوبية، حيث أدى إلى رجوع الناسور، مما جعل التدخل الجراحي مرة ثانية لا يستمر بالناسور بالكامل بعد ثلاثة أشهر من العملية الأولى أمر ضروري.

الاستنتاج: أن وجود حقول صديقة في مكث ناسوري كان مشابهة عرضة محددة، يعتبر مؤشرا لمكثة حدوت تغييرات غير مفيدة، وهذا ييج بإجراء العملية الجراحية في وقت مبكر أمرًا ضروريًا. كما ويجب نقاش تشخيص وعلاج الأفلا في ضوء نشأة الجانيين أمر مستحب.