Case Report

Fourth branchial cleft cyst – case report

Cisto branquial da 4ª fenda – relato de caso

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ABSTRACT: Introduction: Branchial cysts are congenital tumors, resulting from embryonic defects that affect the branchial arches. Congenital cervical abnormalities are usually diagnosed in the first years of life. Objective: To inform, discuss and analyze treatment approaches for this type of congenital cyst. Method: Case report and analysis of data, diagnoses and approaches based on the literature addressing Fourth Branchial Cleft Cysts. Results: Cysts can manifest late, but fistulas are almost always diagnosed at birth or in childhood. They are extremely rare: it is estimated that 95% of branchial cyst anomalies involve the second cleft; of the remaining 5%, almost all arise from the first and third clefts. There are about 45 cases of fourth cleft cysts reported in the literature. The diagnosis is primarily clinical, but the ultrasound can be used for the differential diagnosis of a branchial cyst. Computed tomography will show air-fluid level in the anterior portion of the neck, in front of the thyroid and trachea, which may compress the trachea, causing respiratory distress in childhood. The treatment of branchial anomalies is surgical excision. A 9-month old female patient was being followed up after conservative treatment of a cervical mass (branchial cyst). The cyst appeared immediately after birth, but there was there was spontaneous drainage of the cyst into the esophagus a few days later. After nine months, the patient returned due to a progressive increase of the lesion, which, after physical examination and imaging exams, led to the diagnosis of a fourth branchial cleft cyst. A surgical procedure was performed to remove the cyst along with the left thyroid lobe (partial thyroidectomy). Conclusion: After the excision of the lesion, the patient made a good recovery. She was then referred to the infirmary and later discharged with outpatient follow-up by a general pediatrician.

Keywords: Branchial anomalies; Branchioma; Branchial region; Congenital tumors; Neck mass.

RESUMO : Introdução: Os cistos branquiais são tumores congênitos laterais, resultantes de defeitos de desenvolvimento embrionário que afetam os arcos branquiais. As anomalias congénitas cervicais são mais comumente diagnosticadas nos primeiros anos de vida. Objetivo: Informar, discutir e analisar condutas para tratamento desse tipo de cisto congênito. Método:...
Relato de caso e análise de dados, diagnósticos e conduta baseada na literatura referente a Cisto Branquial da 4ª Fenda. Resultados: Os cistos podem se manifestar tardiamente, mas as fistulas são, quase sempre, diagnosticadas ao nascimento ou na infância. São extremamente raros, estima-se que 95% das anomalias das fendas branquiais sejam da 2ª fenda; das 5% restantes, quase todas são da 1ª ou 3ª fenda. O diagnóstico é principalmente clínico, mas a ultrassonografia pode auxiliar no diagnóstico diferencial de um cisto branquial. O tratamento das anomalias branquiais é a excisão cirúrgica. Lactente sexo feminino, 9 meses de idade em acompanhamento de cisto branquial com conduta conservadora. O surgimento da massa se deu logo ao nascimento, havendo drenagem espontânea do cisto para o esôfago alguns dias depois. Apesar de um acompanhamento de câncer devido a aumento progressivo da lesão que correlacionando com exame físico, exames de imagem levaram ao diagnóstico de cisto de 4ª fenda branquial. Realizada cirurgia para remoção de Cisto juntamente com retirada de lobo esquerdo da tireoide (tireoidectomia parcial). Conclusão: Após a resolução da lesão paciente evoluiu satisfatoriamente sendo encaminhado para enfermaria e posteriormente alta com acompanhamento ambulatorial com pediatra geral.

Palavras-chaves: Anomalias branquiais; Cisto branquial; Fendas branquiais; Tumores congênitos; Massa cervical.

INTRODUCTION

Branchial or bronchogenic cysts are always located in the lateral region of the neck, and, for this reason, many authors still call them lateral cervical cysts. They appear at the anterior border of the sternocleidomastoid muscle in its upper third. When cysts grow to large sizes, they occupy the anterior surface of the sternocleidomastoid muscle, invading neighboring areas such as the suprahoid and infrahyoid muscles, the parotid gland, and deeper regions such as the lateral pharyngeal space, which frequently has a closer connection with the cyst through a small opening common in branchial fistulas. More than 50% of neck masses that require surgical therapy are congenital and present as cystic lesions. The two most common congenital neck masses are the branchial cyst and the thyroglossal duct cyst.

OBJECTIVE

To inform, discuss and analyze treatment approaches for this type of congenital cyst.

METHOD

Case report of a fourth branchial cleft cyst.

CLINICAL CASE DESCRIPTION

A 12-day-old patient called M.J., weighing 3.5Kg, female, was taken into the hospital due to a “lump” in the left cervical region, noticed by the parents. In the anamnesis, the main complaint was the sudden appearance of a “lump in the neck”. According to the parents, the cervical mass located in the left submandibular region had appeared 4 days earlier, was painless and had no signs of inflammation, fever, or progressive increase. They denied fever or any other symptoms. Pregnancy had no complications, and the child was exclusively breastfed.

On physical examination, the child was awake, reactive to handling and interacting with the examiner. Muscle tone and motor response were present, and reflexes were symmetrical. Pupils were medium sized, isochoric, and reactive to light. General examination showed that the patient was hydrated, had normal color, and had no cyanosis or jaundice. It also showed good peripheral perfusion, eupnea and lymphadenomegaly in the submandibular chain, fixed on the left side, with no pain or signs of inflammation or fever. Cardiovascular exam revealed regular rhythm, in two beats, with no murmur. Respiratory auscultation revealed vesicular murmur, with no adventitious sounds. The abdomen was flat, flaccid, had normal peristalsis and no visceromegaly. Lower limbs had no edema and pulses were palpable and broad. Orofaringe exam and otoscopy showed no alterations.

After the anamnesis and physical examination, an evaluation by the hospital’s pediatric surgery service was requested. After the evaluation of the pediatric surgery, the diagnosis of Branchial Cyst was suggested. For diagnosis confirmation, the following exams were requested: cervical ultrasound, cranial ultrasound, computed tomography of the cervical spine, magnetic resonance of the neck, chest radiography and blood count.

Cervical ultrasound showed: non-individualized left submandibular gland; thyroid gland: non-individualized left lobe, with image of a cyst with a thick capsule, filled with fluid, predominantly echogenic, mobile when changing position, with peripheral flow on color Doppler. Isthmus = 0.2 cm.

Neck CT: hypoattenuating and expansive cystic lesion, well-delimited, measuring 3.2 cm, located in the left lateral cervical region, determining hypopharynx/contralateral tracheal deviation. Report suggested Branchial Cleft Cyst.

Neck MRI showed an expansive cystic lesion, measuring about 3.4 x 2.6 cm, in the region of the thyroid on the left side, with contralateral tracheal deviation. The appearance is consistent with a congenital cystic lesion, suggesting Branchial Cyst.

With the anamnesis, physical examination, and the results of the exams, the diagnosis of Branchial Cyst was suggested. The treatment approach was conservative at the time, as there was spontaneous drainage of the cyst into the esophagus. The patient was discharged and referred for outpatient follow-up to evaluate the need for future
After nine months, the child returns to the service, now weighing 8,400 kg, with a complaint of growth of the neck mass on the left side, starting a few days earlier, associated with intense crying and irritability. On physical examination, the patient was irritable and crying. General examination showed the presence of a hard nodule, painful on palpation, in the left cervical region, with no signs of inflammation or fever. Complete blood count, urine sediment and culture and left cervical ultrasound were requested. Later, cyst puncture and culture of the material were performed, resulting in positive culture for Klebsiella Pneumoniae.

The blood count showed leukocytosis. Cervical ultrasound showed a cystic formation with thick and homogeneous content, regular contours, and thin walls, measuring 2.1 x 2.6 cm, in the left anterior cervical region, with apparent communication with the left thyroid lobe.

The neck CT demonstrated total closure of the airway between larynx and trachea at that moment.

The set of findings of the clinical history, physical examination and imaging tests corresponded to the diagnosis of fourth branchial cleft cyst. Then, a surgical procedure was performed to remove the fourth branchial cleft cyst along with the left thyroid lobe (partial thyroidectomy).

The material collected during the surgery was sent for biopsy. Anatomopathological analysis of the left thyroid lobe was performed and returned the following results: macroscopic examination showed an elliptical fragment of skin and subcutaneous tissue measuring 2.0 x 0.5 cm, with a striated and light brown surface, and an irregular fragment of grayish tissue with a soft consistency, measuring 3.0 cm. Cuts were firm, with whitish areas. Microscopic examination showed: thyroglossal cyst, with significant chronic inflammatory infiltrate; granulomatous reaction of the foreign body type; thyroid parenchyma, with interstitial fibrosis; slightly thickened epidermal walls; significant fibrosis of the dermis.

After surgery, the patient was referred to the Pediatric Intensive Care Unit, and made a good recovery. She was then transferred to the infirmary and discharged a few days later. Postoperative management consisted of clinical observation, with scheduled visits to a pediatrician for evaluation.
DISCUSSION

The complex anatomy of the neck favors the emergence of several types of congenital anomalies, which must be differentiated from inflammatory diseases and neoplasms. As in other regions, congenital cervical anomalies are usually diagnosed in the first years of life\(^1\). There are several cervical tumor-like lesions that can appear in the cervical region of children or young adults\(^3\).

The physical examination must be complete to rule out the possibility of other anomalies in different regions of the body\(^4\). Palpation and auscultation of cervical tumors, when indicated, are important for evaluating mobility, inflammatory signs, and vascular flow. These measures can help preventing accidents such as inadvertent puncture of vascular tumors, which can lead to the formation of hematomas\(^4\).

Auxiliary diagnostic methods include ultrasound, computed tomography, and fine needle aspiration. The ultrasound can determine if the lesion is solid or cystic. Its use in conjunction with needle aspiration is important for selecting cases that should or should not be punctured and locating the best site for puncture\(^4,\).

Computed tomography is only used when physical examination and ultrasound are not conclusive or when the dimensions of the lesion indicate the need for a better assessment of the deep planes of the neck and the anatomical relationships between the lesion and important structures\(^4\).

Branchial cysts are congenital tumors resulting from embryonic defects that affect the branchial arches\(^4,\).

They present as cysts or fistulas that are usually congenital, but can manifest throughout life. Cysts may manifest late, but fistulas are almost always diagnosed at
birth or in childhood. The diagnosis is primarily clinical, but the ultrasound can be used for the differential diagnosis of a branchial cyst.

Fourth branchial arch anomalies are considered a theoretical possibility, despite the existence of case reports. These anomalies would descend towards the thorax, developing in the neck after passing under the aorta or under the subclavian artery, and the internal orifice would be in the cervical esophagus. They are extremely rare: it is estimated that 95% of branchial cleft anomalies involve the second cleft; of the remaining 5%, almost all arise from the first and third clefts. There are about 45 cases of fourth cleft cysts reported in the literature.

Its presentation can range from a cystic mass in the lower part of the neck, in the suprasternal notch, to a neck mass associated with acute respiratory distress in the perinatal age.

Computed tomography will show air-fluid level in the anterior portion of the neck, in front of the thyroid and trachea, which may compress the trachea, causing respiratory distress in childhood. The treatment of branchial anomalies is surgical excision. Care measures and complications depend on the anatomical relationships of each lesion. The surgery can be performed endoscopically, to cauterize the orifice next to the piriform sinus when the sinus is small, or externally, when the sinus is bigger.

The origin of lateral cervical cysts still remains unclear, with four theories being proposed for their appearance, including incomplete obliteration of the gill cleft mucosa, which will remain dormant until a later period of life and expands due to some external stimulus. It is also postulated that the cysts could represent vestiges of the precervical sinus or even that the lateral cervical cysts originate from the thyropharyngeal pouch. However, the most accepted and widely discussed theory relates the origin of cysts to cystic degeneration of lymphoid tissue.

As in other regions, congenital cervical anomalies are usually diagnosed in the first few years of life.

CONCLUSION

The importance of this case report is due to the rarity of fourth branchial cleft cysts reported in the literature.


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