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Lemierre's Syndrome in Pediatric Patients: Case Report

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Abstract **Case Report**

Introduction: Lemierre's syndrome is a severe and extremely rare septic thromboembolic event of an infection usually located in the neck and head. It is a rapidly progressive disease that can cause fatal consequences when there is no timely treatment. ***Clinical Case:*** A 6-year 5-month old female patient was evaluated due to a clinical picture of a continuous burning pain in the left anterior cervical region of moderate intensity of 3 weeks of evolution that worsens with a sudden mass growth at the mentioned level. Physical examination revealed an asymmetric neck with limited mobility due to the presence of an oval mass of approximately 6 cm in diameter located in the left anterior cervical quadrant. A CT scan showed an invasive process in the left infrahyoid region that is enhanced with the use of intravenous contrast medium, measuring 4.7 cm x 3.8 cm x 4.2 cm, with an approximate volume of 39 ml. In addition, a hypodense image is observed in the left internal jugular vein suggesting thrombosis. Drainage of a cervical abscess is performed, revealing a mass in the left external segment. The result of the study of the collected purulent fluid reports isolation of two types of germs: Bacteroides fragilis and Peptococus species. Considering also the radiological finding of the internal jugular vein thrombosis, anticoagulation with low molecular weight heparins was started. Patient with good evolution of clinical picture. ***Conclusions:*** Lemierre's Syndrome is a disease in which early diagnosis and treatment can reduce morbidity and mortality, especially in pediatric patients, where its incidence is extremely rare. However, specific clinical data can guide us to its diagnosis.

Keywords: Neck mass, Lemierre's Syndrome, Jugular vein thrombosis.

INTRODUCTION

Lemierre's syndrome is a severe and extremely rare septic thromboembolic event of an infection usually located in the neck and head [1]. When the peripheral location origin can be categorized as "atypical." It was first described by André in 1936 as a post-anginal septicemia, in which the isolated germ was Fusobacterium necrophorumy [1]. It is a rapidly progressive disease that can cause fatal consequences when there is no timely treatment.

CLINICAL CASE

6-year 5-month old female patient with adequate physical and psychomotor development, with complete vaccination schedule for her age, father diagnosed with melanoma, with a relevant background of bacterial pneumonia and febrile seizures at 2 years of age without neurological sequelae, bacterial thyroiditis at the age of 5 treated with intravenous antibiotic scheme, in addition to incidental finding of TIRADS IV thyroid nodule where fine needle puncture (FNA) was planned, but not performed due to abandonment of treatment by the patient. Within the surgical aspect: Resolved right inguinal herniorrhaphy at 3 years of age; no known allergies so far and no reference to travel to endemic and/or tropical areas in the last month.

She was evaluated due to a clinical picture of a continuous burning pain in the left anterior cervical region of moderate intensity of 3 weeks of evolution that worsens with a sudden mass growth at the mentioned level, this intensifies the pain that is accompanied by odynophagia and rhinorrhea.

Physical examination revealed teeth in general in poor hygienic conditions, asymmetric neck with limited mobility due to the presence of an oval, fixed, hard consistency, irregular edges, hot and painful on palpation mass of approximately 6 cm in diameter, located in the left anterior cervical quadrant, with a bright halo in the center and erythema around the circumference.

Figure 1: Mass in the left anterior cervical quadrant

It is complemented with laboratory and imaging studies with evidence of inflammatory response: leukocytes 15880 K/ul [4.50 - 12.00], neutrophils 1230 K/ul [1.4 - 7.0] in addition to elevated acute phase reactants with an erythrocyte sedimentation rate (ESR) of 52 mm/h [0 - 15], C-reactive protein 12.93 mg/dl [0.00 - 0.50] and procalcitonin 0.65 ng/ml. Similarly, in the neck ultrasound scan, there is evidence of heterogeneous image measuring 4.3 cm x 2.6 cm x 4 cm, with a volume of approximately 35 ml, that produces right displacement of midline structures and in a caudal direction to the carotid artery, with peripheral vascularization. The carotid artery Doppler study shows permeable flow and in the internal jugular vein there is a hyperechoic image compatible with chronic thrombus, additional finding of reactive lymph nodes in the left IIB level, with integrity of muscular layers.

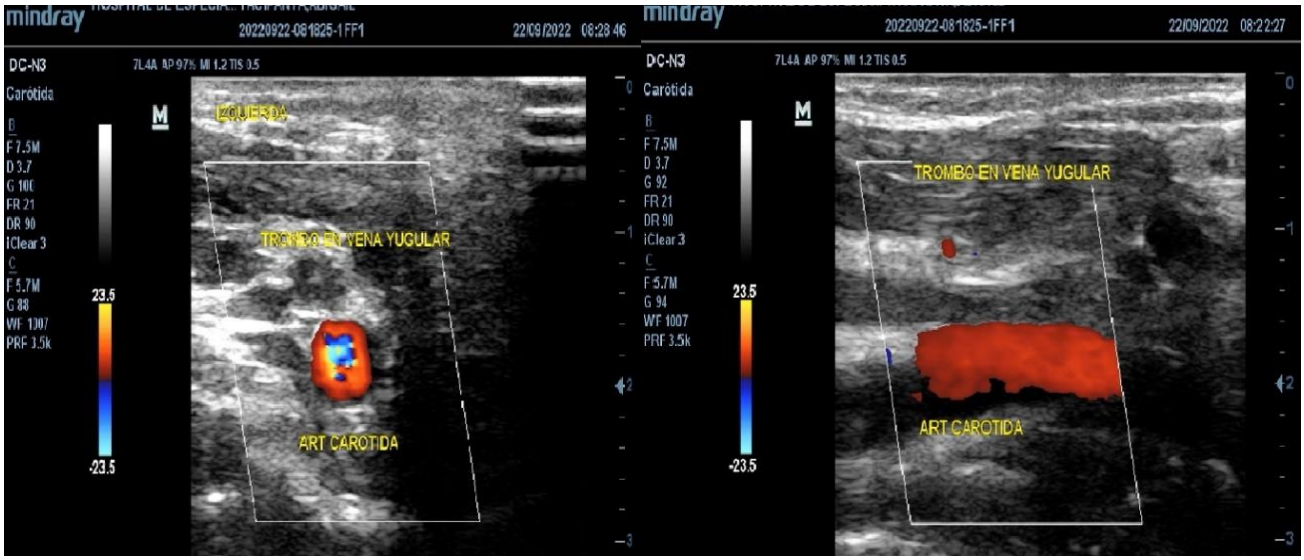

Figure 2: Ultrasound images showing the Left Carotid Artery with permeable flow, presence of hyperechoic image in Left Internal Jugular Vein compatible with chronic thrombus

The simple and contrast-enhanced neck CT scan showed an invasive process of thick capsule with multiple septa in the left infrahyoid region that is enhanced with the use of intravenous contrast medium, measuring 4.7 cm x 3.8 cm x 4. 2 cm, with an approximate volume of 39 ml. However, the liquid content is approximately 25 ml; this produces right displacement of midline structures, of anterior location to carotid artery and external to sternocleidomastoid muscle, extends to left prevertebral spaces without causing compression of the esophagus. Also, the left cervical lymph nodes are enhanced with the use of contrast medium which leads us to an inflammatory process. The thrombus in the left internal jugular vein from the level of the common carotid artery bifurcation to its innominate end.

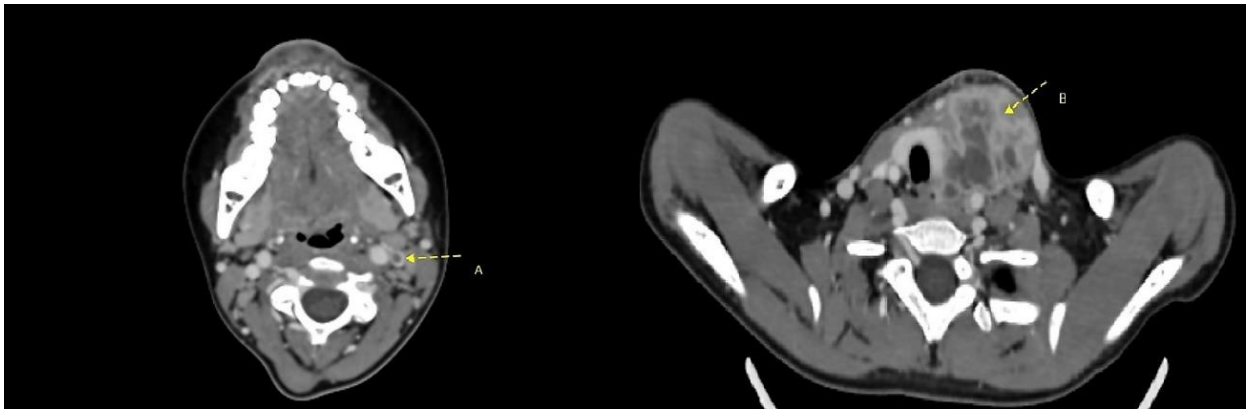

Figure 3: A: Hypodense image in the left internal jugular vein from the level of the common carotid artery bifurcation to its innominate end suggesting thrombosis, B: Image suggesting an intrusive process in the left infrahyoid region

It was decided jointly, between the clinical and surgical departments, to start a combined empirical intravenous antibiotic scheme for cervical abscess based on Clindamycin 200 mg every 8 hours and Ceftriaxone 800 mg every 12 hours, in addition to drainage of the abscess.

Under asepsis and antisepsis standards prior to signing the informed consent, with hemodynamic stability and acceptable infectious parameters, the cervical abscess drainage is performed, revealing a mass in the left external segment from which purulent fluid is extracted (35 ml) from which samples are taken for microbiological culture, a 1cm thick capsule is also identified, which is also submitted for study. The result of the study of the collected purulent fluid reports isolation of two types of germs: Bacteroides Fragilis and Peptococus Species, and the histopathological analysis describes fibrotic tissue with mononuclear aggregates, polymorphonuclear type infiltrate with predominance of neutrophils and eosinophils confirming mass and liquid characteristics compatible with abscess.

Considering also the radiological finding of the internal jugular vein thrombosis, anticoagulation with low molecular weight heparin at a dose of 1 mg/kg was started 24 hours after the immediate postoperative treatment. The patient underwent 4 days of postoperative hospital monitoring with baseline water intake, optimized analgesia with adequate pain tolerance, antibiotic regimen for 5 days and anticoagulation. Control of infectious laboratory parameters in decline was performed: leukocytes 8260 K/UL, neutrophils 3700 K/UL, ESR 50 MM/H, CRP 2.59 MG/DL and control ultrasound study 7 days after the previous one showed an ill-defined hypoechoic image in the surgical area that could correspond to a scarring process and in relation to the internal jugular vein. A 30% recanalization of the vessel lumen is observed, so in the context of clinical, analytical and radiological improvement, hospital discharge is indicated with prescription of rivaroxaban 5 mg at least every 12 hours to continue joint outpatient treatment.

DISCUSSION

Septic thrombophlebitis of the ipsilateral internal jugular vein may cause septic emboli. It occurs after a bacterial infection located in the head or neck, which may be a pharyngotonsillitis or an abscess. This clinical picture is called Lemierre's Syndrome [2]. This is a serious situation that can quickly become complicated without treatment, causing sepsis, and carries a high risk of morbidity and mortality, mainly due to delays in the diagnosis, due to the lack of knowledge on the disease [2]. Its onset usually occurs in patients with no medical history and between 15 and 30 years of age, being exceptional the cases in pediatric patients [1].

Its prevalence is 1 to 10 cases per million person-years [3] and the mortality rate is 4% to 9%; when there is mediastinal extension, mortality rises to 40%, although it has been decreasing due to the appearance of powerful broad- spectrum antibiotics. The male gender has a higher incidence than the female one in a ratio of 2:1. [1].

The germ mainly implicated is the gram-negative anaerobic microorganism Fusebacterium spp. [2, 4]. The F. necrophorum species is the most commonly implicated, followed by F. nucleatum [1]. This microorganism is part of the bacterial flora of the oral cavity, gastrointestinal tract and female reproductive system [2]. However, the presence of Fusobacterium spp. is not indispensable, because it is a difficult microorganism to isolate, even more, when antibiotic therapy has already been initiated. There are other bacteria that can cause Lemierre's syndrome such as Pseudomona aeruginosa, Staphylococcus spp, Streptococcus and Bacteroides spp, the latter isolated in the case of our patient [1, 2].

Its pathophysiology is still uncertain, however, there is a theory that the weakening of the mucous membranes produces proliferation of this microorganism which reaches the bloodstream and generates septic emboli [2]. Dissemination develops from the initial focus to the intravascular space through the tonsillar and peritonsillar veins and finally the possibility of lymphatic dissemination has been raised [5].

Clinically, the symptoms presented may be fever, pain and edema of the neck, generally after the diagnosis of an infection located in the neck region [6]. When septicemia occurs, due to the anatomical relation, the first symptoms to appear are of pulmonary origin. Internal jugular vein thrombosis does not usually show symptomatology, it is essential the support of imaging tests to confirm its finding, however, its presence may be the first symptom of a malignant disease and the study of the cause must be detailed [7].

The contrast-enhanced CT scan continues to be the most used exam and with better results, intraluminal filling defects and enhancement of the affected jugular vein will be observed in addition to soft tissue swelling [1]. Complications such as pulmonary embolism, osteomyelitis and abscesses are also observed by this test. Doppler ultrasound can be used as it is a less invasive test, although the results are not always positive as it is an operator- dependent test [4]. Other diagnostic tests that can be used are oropharyngeal and hematology cultures to rule out the presence of septicemia.

Regarding treatment, management should be multidisciplinary and inpatient, without delaying the initiation of broad-spectrum antibiotics plus early surgical drainage [1]. The antibiotic scheme is not established due to the possibility of beta-lactamase producing strains [2], for this reason it should be adapted to the results of the antibiogram. Internal jugular vein ligation is only reserved when there is persistent embolism in septic patients receiving optimal antibiotic treatment, severe uncontrolled sepsis or extensive septic thrombosis [1, 3]. The use of anticoagulation is controversial due to the risk of hemorrhage in pediatric patients. The benefits of these are prevention of new thromboembolic events, and also prevent the thrombosis from extending. It can be used up to 3 months according to the recommendations of the clinical guidelines for provoked venous thrombosis. The agent and dose will depend on the clinical characteristics of each patient focused on reducing the risk of hemorrhage [7, 8].

CONCLUSION

Lemierre's Syndrome is a disease in which early diagnosis and treatment can reduce morbidity and mortality, especially in pediatric patients, where its incidence is extremely rare. However, specific clinical data can guide us to its diagnosis.

As in the above case, the initial treatment with antibiotic therapy and early drainage of the cervical mass helps to prevent complications that may be irreversible.

Due to the limited literature on Lemierre's syndrome, we recommend continuing with the study of related cases.

CONFLICT OF INTEREST

We, the authors, declare that we have no personal, financial, intellectual, economic, and corporate conflicts of interest.

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