CASE REPORT

Nephrotic syndrome in a pediatric patient with paraganglioma. Case report

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ABSTRACT

Introduction: nephrotic syndrome is due to diverse etiologies; only a relatively small number of diseases are responsible for the majority of patients seen.
Patient information: a 14-year-old African female patient is presented with previous health history until detecting a left cervical enlargement, with slow growth of two years of evolution. Generalized edema was noted until the diagnosis of nephrotic syndrome was confirmed, which was associated with a carotid body tumor (paraganglioma) and acute renal dysfunction. The patient required hemodialysis treatment according to needs without functional recovery and permanent substitution therapies.
Conclusions: It is remarkable the infrequency of this tumor process and its association with a renal lesion that evolved to chronicity in a pediatric patient.
Key words: nephrotic syndrome; cervical paraganglioma

RESUMEN

Introducción: el síndrome nefrótico obedece a etiologías diversas; solo un número relativamente pequeño de enfermedades son responsables de la mayoría de los pacientes atendidos.
Información del paciente: se presenta una paciente africana de 14 años de edad con antecedentes de salud previa hasta detectar un aumento de volumen cervical izquierdo, con crecimiento lento de dos años de evolución. Se constató edema generalizado hasta que se confirmó el diagnóstico de síndrome nefrótico, el que se asoció a un tumor de cuerpo carotídeo (paraganglioma) y a disfunción renal aguda. La paciente requirió tratamiento con hemodiálisis según necesidades sin recuperación funcional y requerimientos de terapias de suplencia permanentes.
Conclusiones: destacan lo infrecuente de este proceso tumoral y su asociación con una lesión renal que evolucionó a la cronicidad en una paciente en la edad pediátrica.
Palabras clave: síndrome nefrótico; paraganglioma cervical

INTRODUCTION

Nephrotic syndrome is due to diverse etiologies; however, only a relatively small number of diseases are responsible for most of the patients seen. The

most common form is idiopathic, in the context of primary glomerular conditions and, less frequently, it occurs secondary to systemic diseases.[1] In the pediatric age it is very rare to find a nephrotic syndrome secondary to tumor processes and, especially, to a paraganglioma which is a tumor of the carotid body, of low incidence, generally benign and of slow growth.[2,3]
The aim of this study is to present a pediatric patient with a carotid body tumor that caused a nephrotic syndrome with renal dysfunction, which required renal replacement therapy with hemodialysis and evolved to chronicity.

PATIENT INFORMATION

A 14-year-old female patient of African origin with a history of health until detecting a tumor process of slow evolution (two years) in the left region of the neck; no study of its etiology had been performed. She went to the Health Services due to decay, anorexia, isolated vomiting and hiccups, facial edema and edema of both feet and decreased urinary volume. She was admitted to the Pediatric Hospital and a referral was made to the Hemodialysis Center of the "Josina Machel" Hospital in Luanda, Angola.
Physical examination revealed pallor of the mucous membranes, infiltrated subcutaneous cellular tissue on the face, on the slopes and on both feet up to the knees (Godet +++), a tumor formation in the left lateral neck region of 3.5x4cm in diameter, slightly movable, slightly painful and hard, and a grade IV/VI systolic murmur. In addition, decreased vesicular murmur in the middle region of the left hemithorax and right base, with crepitant rales and pulmonary silence towards the left base. Respiratory rate was 20 breaths per minute, heart rate 112 beats x minute and blood pressure 90/50mm/Hg. The abdomen was globular with slightly painful hepatomegaly on palpation and positive Tarral maneuver.
Laboratory studies:

- Hemogram
- Hemoglobin: 7g/dl
- Hematocrit: 027vol%.
- Leukogram: $8.3x10^9$/l, Polymorphonuclear 87
- Platelets: $154x10^9$/l
- Hemochemistry
- Creatinine: 11,82 micromol/l
- Urea: 37mg/dl
- Total protein: 3.9g/l
- Albumin: 0,2g/l
- Cholesterol: 657mmol/l
- Triacylglycerides: 148 mmol/L
- Glycemia: 103 mmol/l
- Urine: cloudy foamy appearance, with protein +++, no red blood cells and leukocytes 2xfield.
- Virology negative for hepatitis B, hepatitis C and human immunodeficiency virus.

Imaging studies:

- Ultrasound of soft parts of the neck: the neck region was explored with soft tissue transducer and towards the palpated lesion; an ovoid mass was observed, complex, predominantly hypoechogenic, with well-defined, regular contours, with small areas of lower echogenicity inside (echolucent), possibly related to areas of cystic degeneration, with central and peripheral vascularization at Doppler, predominantly peripheral, measuring approximately 10.5x8.0cm and located in the left superolateral region of the neck, in the thickness of the muscular plane, in intimate contact with the submaxillary gland and anterior to the jugular vein, without infiltrating these structures.
- Computerized axial tomography (CT) of the neck: a solid nodular mass measuring 111x85mm, with a density approximating the soft tissues, was observed at the level of the jugulodigastric space. After the application of contrast showed an agile uptake pattern and a poorly uptaken central area. The referred process marked the deviation of the internal carotid artery and adjacent structures (submandibular gland, parotid and sternocleidomastoid muscle); no bilateral cervical adenomegaly.
- Chest CT: left medium-sized pleural effusion. The parenchymal evaluation showed multiple nodules distributed in the central and subpleural regions in both lung fields, all of them with suspension character (metastatic lesions). In the posterior basal segment of the lower lobe of the right lung a focus of infammatory condensation was identified suggesting an evolving infectious lesion, as in the posterior basal segment of the lower lobe of the left lung. In the mediastinal area there were precarinal and pretracheal adenomegalies and in the aortopulmonary window, the largest towards this last space measured 13mm. Cardiovascular structures were of normal caliber.
- Abdominal CT: enlarged liver with regular contrast uptake without visualization of solid nodular lesions in the parenchyma. Kidneys in usual topography, normal size, with homogeneous and symmetrical contrast uptake. Rest of the intra-abdominal structures without alterations and presence of free fluid in the cavity.

Cytological study:

Fine needle aspiration cytology of cervical lymph node showing fibrous tissue, adipose muscle and salivary gland, in which a focus of granuloma with many foreign body-like giant cells is found. Oval or polygonal cells (chief cells) with uniform or pleomorphic, vesicular or hyperchromatic nuclei arranged in organoid nests surrounded by a vascular stroma (Zellballen) that appeared to correspond to a paraganglioma.

The patient was evaluated by the Nephrology Specialist; his conclusion was clinically consistent with a nephrotic syndrome and a secondary parenchymal acute kidney injury associated with a neck tumor with pulmonary metastasis. It was not possible to perform a renal biopsy and the degree of kidney involvement was K-DIGO 3, without urinary volume.

Therapeutic intervention:

- Hyposodic, normoproteic, hypocaloric diet, 60 to 65% of them, less than 30% of lipid intake, preferably of vegetable origin, with less than 300mg of cholesterol per day.
- A double lumen catheter without tunneling was placed in the left femoral vein (9Fr 11cm catheter).
- Emergency hemodialysis was prescribed for a period of two hours with F5 dialyzer, blood flow of 130ml/minute, ultrafiltration according to weight gain and hemodynamic stability with Na: 140meq/L and temperature of 35.5°C, and anticoagulation with conventional heparin.
- 20% human albumin at the beginning of hemodialysis.
- Hydroelectrolyte balance and urinary volume evaluation. Oral replenishment according to discharge with the intention of maintaining a negative balance.
- Periodic evaluation in joint follow-up with Pediatric and Nephrology Specialists and monitoring of renal function every 24 hours.
- Interconsultation with the Surgery and Oncology Specialists to evaluate the conduct for the neck tumor process.

DISCUSSION

Nephrotic syndrome presents with a high incidence in pediatrics, 2-7 cases per 100,000 children per year.[4,5] It is a clinical-humoral syndrome characterized by the presence of massive proteinuria (higher than 40mg/m^2/h, approximately 1g/m^2/day, or the ratio proteinuria/creatinine in random urine higher than 200mg/mmol, with plasma albumin lower than 25g/l, edema and hypercholesterolemia higher than 250mg/dl or 5.69mmol/l).[5,6,7] The etiology secondary to systemic diseases or some identifiable process causing the glomerular lesion, as neoplastic processes, is infrequent in childhood and the literature is very scarce associated to paragangliomas.

Paraganglioma is a generic designation that, globally, is used to name the family of neuroendocrine neoplasms, which are rare and can originate in the adrenal medulla or in the paraganglia of the diffuse neuroendocrine system. In particular, this term is usually reserved for tumors arising from extra-adrenal tissue.[8] They account for 0.6% of head and neck tumors[2,3,9,10] and may have sporadic or familial presentation.[2,3,10] No environmental, dietary or lifestyle risk factors have been linked to the presentation of paraganglioma.[3,8]

Sporadic cases are more common in females; familial cases occur in 10%, are transmitted in an autosomal dominant manner through the 11q23 locus gene and have a high incidence of presenting bilaterally. The incidence of bilaterality of carotid body paraganglioma ranges from 3 to 8% in sporadic cases and 30 to 33% in familial cases. Seventy-nine percent of head and neck paragangliomas have shown a mutation in the D subunit of the SDH gene, which has suggested the presence of multiple paragangliomas.[3,10]

Up to 41% of cases present identifiable mutations, most of these are hereditary and are associated with a high risk of malignant transformation.[3,11]

Carotid paragangliomas are generally benign tumors, with a low malignant potential, less than 5%;[3,12] however, they exhibit a high morbidity due to

mass effect, besides being the only disease affecting the carotid body and represent 0.03% of all neoplasms.[3,10]

It has been observed that its incidence increases proportionally with altitude due to the fact that the hypoxic stimulus induces hyperplasia in the carotid body.[2,10,12]

The diagnosis of these tumors is often late, patients are frequently asymptomatic, and their growth pattern is slow, so they may present decades before the patient consults.[2] Diagnosis and early surgical resection of the tumor are important, since when they are small they are poorly adherent and easily extirpated, with a lower incidence of complications.[13]

This patient remained hospitalized with follow-up in the Nephrology Service for more than three months, without recovery of renal and therapeutic function and with hemodialysis. She was incorporated to the conventional dialysis program and was followed up by the Oncology Specialist.

The infrequent nature of this tumor process and its association with a renal lesion that evolved to chronicity in a pediatric patient stand out.

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CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.