2022, Volume 9, ID 599 DOI: <u>10.15342/ijms.2022.599</u>

CASE REPORT

Cerebro-Meningeal Hemorrhage Revealing a Pheochromocytoma in a Child

Abdelhak EL Khadi ^a, Youssef Motiaa ^b, Mohammed Aabdi ^b, Hicham Sbai ^b, Smael Labib ^b ^a Intensive care unit, Mohamed V hospital, Faculty of Medicine and Pharmacy, Universit of Abdelmalek Essaadi, Tangier, Morocco ^b Faculty of Medicine and Pharmacy- tangier, Universit of Abdelmalek Essaadi, Tangier, Morocco

ABSTRACT

Pheochromocytoma is a sporadic tumor of the adrenal medulla in the children population. With very polymorphic symptomatology, sometimes causing a diagnosis delay can be revealed in some cases by a complication. We report the case of a child admitted for cerebral-meningeal hemorrhage caused by a pheochromocytoma which was confirmed by a CT scan and urinary dosage of catecholamines.

KEYWORDS: Cerebro-Meningeal Hemorrhage, Pheochromocytoma, Hypertension, Child.

Correspondence: Dr. Abdelhak EL Khadi, Address: Intensive care unit, Mohamed V Hospital, Tangier, morocco. Email: <u>elkhadiabdelhak@gmail.com</u>

Copyright © **2022 EL Khadi A et al.** This is an open access article distributed under the <u>Creative Commons Attribution 4.0</u> <u>International</u>, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

INTRODUCTION

Pheochromocytoma is a catecholamine-secreting tumor of the adrenal medulla, representing 0.1 to 1% of the causes of hypertension [1]. Adults are the most affected, whereas it is rarer in the pediatric population, representing less than 20% of all pheochromocytomas [2]. The clinical symptoms are very polymorphic and even atypical, causing a delay in diagnosis. Pheochromocytoma can evolve into a lethal or even malignant form in 10 to 20% of cases [1].

We report the case of a 9-year-old child diagnosed with pheochromocytoma following a cerebral-meningeal hemorrhage.

CASE REPORT

A 9-year-old child with no medical record was admitted to the ICU to manage an apyretic disorder of consciousness preceded. According to the mother, morning vomiting in jet form with headaches resistant to treatment for two weeks.

The admission examination found an unconscious child with a Glasgow score of 10 with equal and reactive pupils without sensory-motor deficits. A blood pressure of 140 /90 mmHg, the respiratory and cardiac rate was 20 cycles/min and 95 beat/min with a saturation of 95% in room air, the capillary glycemia was correct, the same for the rest of the biological check-up, the rest of the clinical examination was normal. A cerebral CT scan was performed, showing a meningeal hemorrhage with correct ventricular flooding (Figure 1). Treated with an emergency external ventricular bypass in the operating room and was then hospitalized in the ICU for further treatment.

During his stay, the child presented very high blood pressure levels, reaching 180mmhg/100mmhg with the installation of painful abdominal distension evaluated by abdominal ultrasound. Objectifying an inter aortocaval retroperitoneal adenopathy completed by an abdominal CT; showing a hyper vascularized retroperitoneal mass measuring 61*51*40 mm, involving the inferior vena cava and the left renal artery suggesting a pheochromocytoma aspect (figure 2). the dosage of urinary catecholamine was conclusive: vanylmandelic acid (VMA) at 162.4 µmol/24 h (N 5 at 35 µmol/24 h) and homovanillic acid (HVA) at 40 µmol/24 h. The child died of septic shock before the surgical procedure.

DISCUSSION

Pheochromocytomas are neuroendocrine tumors derived from the neural crest and develop at the expense of chromaffin cells located in the adrenal medulla. Extraadrenal localization is possibly called paragangliomas, can grow in diffuse chromaffin tissues. [1] It represents 0.1 to 1% of the causes of hypertension. It is predominant in adults and exceptional in children (less than 20% of all pheochromocytomas) [2]. The age range varies between less than one year and 14 years [3]. With a male predominance and a familial notion in 10% of cases [4].

The clinical symptomatology is very polymorphic due essentially to the synthesis and excessive releasee of catecholamines, in particular norepinephrine and epinephrine, with increased activation of the sympathetic nervous system[5] the diagnosis of pheochromocytoma is often tricky, suspected in front of clinical symptomatology translating the hypersecretion of catecholamine, whereas in 25% of the cases the tumor is of fortuitous discovery during an abdominal radiological examination (CT or MRI) the classical symptomatology includes the clinical triad of Menard; headache, pulse, and sweating associated with arterial hypertension [6] but other clinical signs may appear in children, notably- asthenia, a polyuria-polydipsic syndrome, metabolic disorders (diabetes, thyrotoxicosis), neurological disorders with anxiety, and sometimes convulsions or even bone lesions in malignant forms [3.7.8]



Figure 1: Cerebral CT scan showing cerebral haemorrhage with right ventricular flooding.



Figure 2: Abdominal CT scan showing an inter aortocaval mass

Arterial hypertension can manifest itself in a paroxysmal way, but permanent systolic-diastolic with more severe complications, especially in children, engage the vital and functional prognosis, namely retinopathy cardiomyopathies or cerebral hemorrhages [9]. This is the case of our patient who was admitted in the stage of a cerebral hemorrhagic complication with ventricular flooding Stroke in children. Young adults account for less than 5% of all strokes [10]. The annual incidence in the 0-14 age group (excluding birth-related strokes, intracranial infections, and post-traumatic strokes) is 2.52 cases per 100,000, the risk of cerebral ischemia being the rarest, estimated at 0.63/100,000/year or even 1.2 cases per 100,000 per year according to other authors [11]. While the risk of bleeding is estimated at 1.89/100,000/year [12] Paraclinical examinations have made the diagnosis of pheochromocytoma easier, particularly radiological explorations and plasma or urinary dosage of free catecholamine. The choice of the radiological examination depends on the situation and availability. These explorations are essential to localize the tumor to know the degree of necrosis and determine the extent of the parenchymal lesions.

Abdominal ultrasound is still the only way to localize an isolated tumor, whereas CT and MRI are currently the examinations of choice. The meta-iodobenzyl-guanidine (MIBG) Iodine 123/131 allows a mapping of the whole body for the detection of multiple tumors or ectopic localization [13]

In our situation, abdominal ultrasound was requested in the presence of an abdominal mass syndrome. It was subsequently supplemented by an abdominal CT scan, which indicated the mass's location, vascularization, and extent. On the biological level, the detection of an excessive quantity of free catecholamines (adrenaline, noradrenaline, dopamine) and their metabolites (vanylmandelic acid, methoxylated derivatives) allows the diagnosis of pheochromocytoma to be made, with a sensitivity of up to 99% for free plasma measurements and approximately 95% for urine measurements. [14] In our situation, VMA and HVA confirmed the diagnosis of pheochromocytoma with very high concentrations. The treatment of pheochromocytomas remains surgical with total removal of the tumor consisting of "gently dissecting the patient from the tumor and not the tumor from the patient." Elective ligation of the adrenal veins before all maneuvers of traction and pressure of the tumor is essential [15].

Surgical management requires obligatory :

pre-medication with antihypertensive drugs, in particular alpha-adrenergic blockers, for 7 to 14 days, to control and stabilize the blood pressure figures to prevent any intraoperative catecholamine release accident while avoiding an excessive lowering, damaging at the myocardial and cerebral level. [16]

Blood pressure monitoring with adequate correction of blood volume [14].

A close and rigorous monitoring of the postoperative follow-up may persist for several days because of the hemodynamic instability.

CONCLUSION

Pheochromocytoma is a rare tumor in the pediatric population, with a very polymorphous clinical picture and even severe complications, including cerebral-meningeal hemorrhage. According to our case, we must also think of pheochromocytoma in front of cerebral-meningeal bleeding in children.

AUTHORS' CONTRIBUTIONS

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly work in Medical Journals of the International Committee of Medical Journal Editors. Indeed, all the authors have actively participated in the redaction, the revision of the manuscript, and provided approval for this final revised version.

PATIENT CONSENT

Written informed consent was obtained from the patient's family for the publication of this case report.

REFERENCES

- Badet C, Mornex R. Phéochromocytomes. Encycl Méd Chir (Éditions Techniques, Paris). Endocrinologie. Nutrition. 10- 015-B-50. 1992.
- Réveillon Y, Daher P, Jan D, Buisson C, Bonnerot V, Martelli H, et al. Pheochromocytoma in children: 15 cases. J Pediatr Surg. 1992 Jul; 27(7): 910-1. DOI: <u>10.1016/0022-3468(92)90396-0</u>
- [3] Dubois R, Chappuis JP. Le phéochromocytome : particularités pédiatriques. Arch Pédiatr. 1997 Dec; 4(12): 1217-25. DOI: <u>10.1016/s0929-693x(97)82613-9</u>
- [4] Levine C, Skimming J, Levine E. Familial pheochromocytomas with unusual associations. J Pediatr Surg. 1992 Apr; 27(4): 447-51. DOI: <u>10.1016/0022-3468(92)90333-3</u>
- [5] Deren M, Lentschener C. Anaesthetic management of surrenalectomy for pheochromocytoma: A controversial issue. Prat Anesth Reanim. 2012 Apr; 16(2): 116–121. DOI: <u>10.1016/j.pratan.2012.02.002</u>
- [6] Plouin PF, Chatellier G, Delahousse M, Rougeot MA, Duclos JM, Pagny JY, et al. Recherche, diagnostic et localisation du pheochromocytome. Presse Med. 1987 Dec 19;16(44):2211–5.
- [7] Delgoffe C, Bretagne MC, Hoeffel JC. Phéochromocytomes bénins et lésions osseuses métaphysaires. Arch Fr Pediatr. 1982 Apr :(4);39:259-61.
- [8] Valade S, Chazerain P, Khanine V, Lazard T, Baudin E, Zizaa JM. Métastases osseuses tardives d'un phéochromocytome. Rev Med Interne. 2010 Nov;31(11):772-5. DOI: <u>10.1016/j.revmed.2010.07.004</u>

COMPETING INTERESTS

The authors declare no competing interests with this case.

ACKNOWLEDGMENTS

None.

FUNDING SOURCES None.

- [9] Arnault-Ouary G, Chatal JF, Charbonnel B. Phéochromocytomes. Rev Prat. 1998 Apr 1.48(7):744-8.
- [10] Gulati S, Kalra V. Stroke in children. Indian J Pediatr. 2003 Aug; 70(8): 639-48. DOI: <u>10.1007/bf02724254</u>
- [11] Castelnau P, Favreau A, Krier C, Barthez MA. Strategie diagnostique dans les accidents vasculaires cerebraux ischemiques de l'enfant. Arch Pediatr. 2005 Sep; 12(9):1433-44. DOI: <u>10.1016/j.arcped.2005.01.033</u>
- [12] Schoenberg BS, Mellinger JF, Schoenberg DG. Cerebrovascular disease in infants and children: a study of incidence, clinical features and survival. Neurology. 1978 Aug; 28(8): 763-8. DOI: 10.1212/wnl.28.8.763
- [13] Abecassis JP, Pariete D, Bonnin A. Imagerie des surrénales. EMC. 1992 :9.
- [14] Pacak K, Eisenhofer G, Ahlman H, Bornstein SR, Gimenez-Roqueplo AP, Grossman AB, et al. Pheochromocytoma: recommendations for clinical practice from the First International Symposium. October 2005. Nat Clin Pract Endocrinol Metab. 2007 Feb; 3(2):92-102. DOI: <u>10.1038/ncpendmet0396</u>
- [15] Newman KD, Ponsky T. The diagnosis and management of endocrine tumours causing hypertension in children. Ann NY Acad Sci. 2002 Sep; 970:155-8. DOI: <u>10.1111/j.1749-6632.2002.tb04421.x</u>
- [16] Lenders JW, Duh QY, Eisenhofer G, Gimenez-Roqueplo AP, Grebe SK, Murad MH, et al. Pheochromocytoma and paraganglioma: An endocrine society clinical practice guideline. J Clin Endocrinol Metab. 2014 Jun; 99(6):1915-42. DOI: <u>10.1210/jc.2014-1498</u>