Case report

LABIA MAJORA HEMANGIOENDOTHELIOMA – CASE REPORT

Zlatan Elek, Zvonko Radosavljevic and Blazo Turkovic

Most vascular tumors comprise haemangiomas and hemangioendotheliomas. The peak of their incidence is usually seen in the fourth decade, whereas only 5-10% of them are seen in children. Infantile haemangioendothelioma is found in newborns and is usually discovered right after birth, whereas in case of infants it can be discovered during the first three months after birth. Haemangioendothelioma is described as seen in various organs: central nervous system, liver, urinary tract, retroperitoneum, lungs and heart. Subcutaneous forms are fairly frequent, but in most instances they represent a cosmetic problem, though in certain cases their localisation and size may be the cause of various functional difficulties.

The paper presents a case of a nine-month-old female patient with a tumor in the region of labia majora that appeared at the age of 2 months, according to what parents said. The patient was adequately preoperatively prepered and was operated on under general endotracheal anaesthesia. Surgical intervention consisted of the excision to the extent of reaching the healthy tissue.

Haemengioendothelioma in the region of labia majora is a rarity in terms of clinical practice. The accurate diagnosis of this tumour can be made by histopathological analysis only. Excision of tumor to the extent of reaching a healthy tissue is the treatment of choice. There are also descriptions of relapses occurring upon extensive excisions. *Acta Medica Medianae 2007;46(4):74-76.*

Key words: *haemangioendothelioma, labia majora, excision*

Pediatric Surgery Health Center in Kosovska Mitrovica

Contact: Zlatan Elek
18/15 John Kennedy Street
38220 Kosovska Mitrovica
Phone: O28 422 251
E-mail: drzelek@gmail.com

Introduction

Infantile hemangioendothelioma is a benign tumor and it is usually discovered at birth in 1/3 all hemangioendotheliomas in children (1). As far as localization is concerned, they mostly take hold of central nervous system, retroperitoneum, urinary tract, lung and heart. As is the case with hemangioma, the subcutaneous localization is frequent, and also can be found on extremities, lips, face and nose.

Very rarely, it is localized on the exterior genitals. Rarely has hemangioendothelioma a malignant potential. Histological appearance is marked by proliferation of large spindle-shaped tumors cell associated with necrosis, cell pleomorphism, frequent mitosis and aggressive infiltrative form.

Clinical presentation is benign (2). The best predictor of tumor behavior is size, localization and histological finding. Hemangioendothelioma is locally invasive, slow-growing tumor.

Calcifications are frequent in infants (3). Diagnosis of hemangioendothelioma is made according to anamnestic data, clinical examination, echosonographic transformation examination. However, precise diagnosis is based on pathohistological finding. Unlike hemangioma, it almost never causes profuse bleeding and thrombocytopenia and does not endanger vital functions.

Surgery is certainly a primary treatment modality; however, the local recurrences have been described after complete excision. Recurrence of disease was described ten years later, after the first surgical intervention. For local recurrences, excision is a therapy of choice (3).

Case report

A nine-month-old girl was admitted to Pediatric Surgery Department because of swelling on the right side of genitals. According to parents' saying, the swelling on her genitals showed up for first time when she was two months old. Thencenforth up to notification to the doctor, the swelling did not show tendency to grow. Immediately after reception into the hospital, the girl was examined clinically, radiogically and echosonographically. Labaratory analyses were also done: complete blood count and coagulation of transformation pointed out hyperechogenic

formation at the right side of the genitalia (4,5). Echosonografic examination of abdomen showed a regular finding without the presence of pathological formations and without increased lymph glands in the small pelvis and abdomen. In addition, radiography of the lungs was done, not showing any pathological formations.

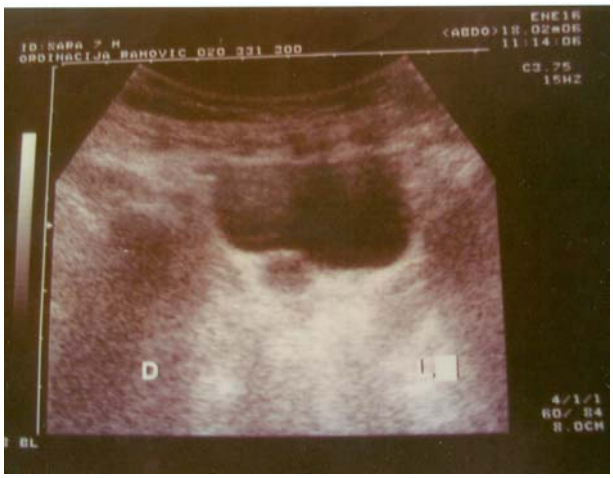

Figure 1. Echosonography of genitalia shows the presence of soft tissue nodule localized in the right labia maior

After an appropriate preoperative preparation, surgical intervention was done including complete excision of the tumor formation. Tumor was red in color, of hard consistency, 2x1,5 cm in size. The operative and postoperative course passed without complications. The wound healed per primam. The stitches were removed seventh days postoperatively.

Figure 2. The patient with tumor in the area of labia majora

Microscopic examination showed the subcutaneous, poorly limited tumor consisting of lobules, bigger and smaller vascular, mainly capillary, vessels containing erythrocytes covered with endothelial cells with poorly expressed atypia and polymorphism without pathological mitosis.

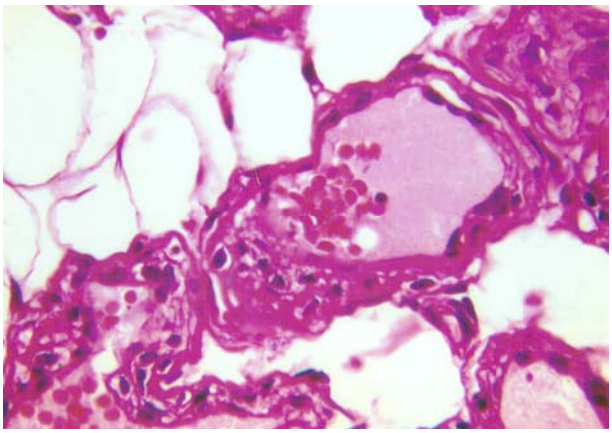

Figure 3. Histological appearence of preparation; vascular lobules marked with endothelial cells dominate

Stroma is very deficient, and there are focal fields of necrosis. Small lobules of tumor tissue are localized in the surrounding fatty tissue. Tumor infiltrations appeared to be disorganized, consisting mostly of cells of different morphological presentation.

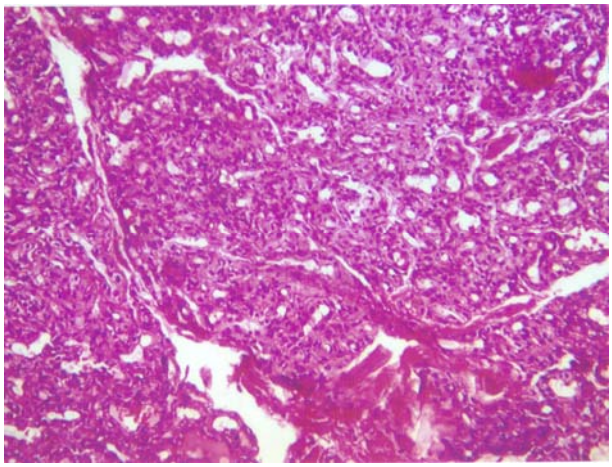

Figure 4. *Histological appearance of the preparation, pronounced ayipia and polymorphism with highly deficient stroma and focal necrosis fields*

The final pathological diagnosis was infantile hemangioendothelioma.

Discussion

In case of infantile hemangioendothelioma, there is usually one tumor nodule, located mostly in the liver, central nervous system, spleen and retroperitoneum. Tumor size varies from 0,5 to 10 cm (5,6). Tumor usually occurs in adults aged 30-40 years old and very rarely in children. Subcutaneous forms have been described so far, but their localization on genitalia is very rare.

The definite diagnosis is made only after pathohistological examination.

There is no other procedure (echosonography, CT) to confirm this diagnosis with certainty. In case of our patient, clinical examination, labora-

tory investigation, radiography of the lungs and echosonographic examination of the change were done because of preoperative preparation. Complete excision of the tumor change was done to the healthy tissue.

The diagnosis was based on pathophystological examination of the excision preparation.

Parmar et al. (7) treated their patients suffering from infantile hemangioendothelioma mostly by undertaking the excision of tumor as a regular mode treatment. The diagnosis was based upon the pathohistological verification.

Intra- and postoperative complications are very rare. Depending on the localization, extensive surgical interventions are undertaken aiming to remove the tumor tissue.

Kitigawa et al. (8) presented a case of infantile hemangioendotheliom affecting the distal phalanx of the second finger on the right hand which was treated by amputation of the phalanx. Fraser et al. reported a case of a patient with ileocaecal haemangioendothelioma treated by the right hemycolectomy.

After the surgical treatment, prognosis is very good.

Conclusion

Infantile haemangioendotheliom is frequent vascular tumor of organs of the central nervous system, liver, spleen and lungs. It is very rarely located on the genitalia. The definite diagnosis is based upon pathohystological examination. Complete healing can be achieved after excision, though recurrences are reported even ten years after excision.

References

1. Cvetković P. Klinicka dečja onkologija. Beograd: 2000. p.255-6.
2. Semina I, Pagella F, Delu G, Tadeschini A, Luieneti O, Zapoli F et al. Endoscopic treatment of ethmoidal haemangioendothelioma: case report and rewiev of the literature. Am J Otorinol 2006;27(4):287-90.
3. Nezir Z, Pervez S. Malignant vascular tumor of liver in neonatus. J Pediatr Surg 2006;41(1):49-51.
4. Vagel AM, Alesbury IM, Fox VL, Fishman SI. Complex pancreatic vascular anomalies in children. J Pediatr Surg 2006;40(3):473-8.
5. Fraser SA, Deschen J, Bloom C, Gonday PH. Ileocekal haemangioendothelioma, report of a case and rewiev of the literature. Dig Colon Rectum 2006; 41(3): 276-9.
6. Maiyadi AV, Brat DI, Devi BI, Mahadevan A, Shankan SK, Sastry KV. Spinal epidural epitheloid haemangioma, case report and rewiev of the literature. Pediatr Neurosurg 2005; 41(3): 155-7.
7. Parmar RC, Bavdekar SB, Borwankar SS, Muranjan MN, Shenoy A. Infantile haemangioendothelioma. Indian J Pediatr 2001; 68(5): 459-61.
8. Kitigawa Y, Ito H, Iketani M, Hirikawa S, Jokojama M, Majeda S. Epitheloid haemengioendothelioma of the falanx a case report. J Hand Surg 2005;30(3):615-9.

HEMANGIOENDOTELIOM VELIKE USNE – PRIKAZ BOLESNIKA

Zlatan Elek, Zvonko Radosavljević i Blažo Turković

Većinu vaskularnih tumora čine hemangiomi i hemangioendoteliomi. Vrh incidence njihovog pojavljivanja je u četvrtoj deceniji a samo 5-10% se nalazi kod dece. Infantilni hemangioendoteliom se otkriva kod novorođenčadi na rođenju, a kod odojčadi se može otkriti u toku prva tri meseca po rođenju. Hemangioendoteliom je opisivan u različitim organima: centralnom nervnom sistemu, urinarnom traktu, retroperitoneumu, plućima i srcu. Subkutane forme su česte i one u većini slučajeva predstavljaju kozmetički problem, mada u određenim slučajevima njihova lokalizacija i veličina može biti uzrok različitih funkcionalnih smetnji. U radu je prikazana bolesnica stara 9 meseci sa tumorom u predelu desne velike usne, koji se javio po kazivanju roditelja u drugom mesecu života. Bolesnica je adekvatno preoperativno pripremljena i operisana u uslovima opšte endotrahealne anestezije. Hirurška intervencija sastojala se u eksciziji tumora do u zdravo tkivo. Hemangioendoteliom velike usne je raritet u kliničkoj praksi. Tačna dijagnoza ovog tumora moguća je samo uz patohistološku verifikaciju. Ekscizija tumora do u zdravo tkivo je tretman izbora. Opisivani su recidivi i posle izdašnih ekscizija. *Acta Medica Medianae 2007; 46(4):74-76.*

Ključne reči: *hemangioendoteliom, velika usna, ekscizija*