Childhood Abrikossoff tumor in a rare localization

Dziecięcy guz Abrikossoffa w rzadkiej lokalizacji

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ABSTRACT

Abrikossoff tumor is an uncommon neoplasm of soft tissue, mostly benign but cases of aggressive malignant form with metastases were also reported. GCT can occur at any area of the body (head and neck are the most common but also limbs and vulva). Local surgical excision with wide margin is a treatment of choice but possibility of recurrence, multiple location and malignancy in time must be considered. This report presents a rare pediatric case with Abrikossoff tumor.

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Key words: neoplasm-benign, surgery, therapy-systemic

STRESZCZENIE

Guz Abrikossoffa jest rzadko występującym nowotworem tkanek miękkich, głównie łagodny, ale również istniały doniesienia o przypadkach agresywnych form z przerzutami. Guz ziarnistokomórkowy (GCT, *granular cel tumor*) może pojawić się w każdej części ciała (głównie na głowie i szyi, ale również na kończynach i sromie). Leczeniem z wyboru jest miejscowe wycięcie z szerokim marginesem, ale musi być brana pod uwagę możliwość wznowy w różnych lokalizacjach i zezłośliwienia. Ten raport przedstawia rzadki, pediatryczny przypadek guza Abrikossoffa.

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Słowa kluczowe: nowotwory łagodne, chirurgia, terapia systemowa

INTRODUCTION

Granular cell tumors (GCTs), or Abrikossoff tumors, are very seldom, usually reported in the literature as single cases, occurring in about 0.02% of human neoplasms and representing 0.5% of all soft tissues [1, 2]. GCTs may occur at any site of dermis or subcutaneous tissue of the body. Head and neck is the most common location especially oral cavity and tongue. Other locations are also reported but more rare [3].

GCTs can affect people at any age, race but are more common in females (M/F ratio = 1:2) [4]. GCTs usually manifest in adults

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in the fourth to sixth decade of life. Occurrence in childhood and adolescence is rare, but some studies show appearance of 12% of all cases [5].

Lesions are usually benign, the malignant variant of GCTs is only 1–2%. Malignant GCT has a poor prognosis with local recurrence over 30%, and metastatic disease present within two years in half of the cases. Treatment of choice is local surgical excisional biopsy with wide margin which is also for diagnosis [6]. Regional steroid therapy combined with surgical treatment may be effective in multifocal and recurrent lesions [7].

In this report, we present a pediatric case with Abrikossoff tumor and the review of actual literature.

CASE REPORT

A 10-year-old girl with no remarkable past medical history. Tumor localized in the left elbow pit was accidentally noticed by parents six months before performing a diagnostic process. Lesion was solitaire, skin — coloured, diameter of 5 mm. Girl did not report any pain, itching, discharge or burning. Parents did not observe enlarging of lesion.

There was no trauma of that area prior to tumor occurrence. Dermatological consultation was conducted but no dermatoscopy and imaging were performed. The general physical examination was normal. Lab tests were within normal limits. Lesion was surgically excised with wide margin of healthy-looking tissue. **Histopathological examination** confirmed diagnosis of Abrikossofff tumor — large cells with granular cytoplasm localized around nerves and dispersed in subcutaneous tissue. Immunohistochemical staining: S — 100 positive. Margin of healthy tissue: 0.5 mm width and 1.5 mm depth. Ultrasonography (US) showed three enlarged lymphatic nodes above mediastinal inlet — the largest 18 × 9 mm, other two: 9 × 4 mm and 5 × 2.5 mm — of decreased echogenicity and hilar pattern of vascularization (to further observation). In US of axillae — single lymph nodes of proper shape and echogenicity, proper image and hilar pattern of vascularization. In US of area of cicatrix post tumor excision — no pathological foundings in soft tissue were described. Patient was discharged from clinic in good general condition to further observation in outpatient clinic of pediatric oncology.

DISCUSSION

This article presents a case report of pediatric granular cell tumor with typical symptoms but with unusual localization. **GCTs may occur at any site of dermis or subcutaneous tissue of the body.** Tumors localized in dermis spread in upper layers of subcutis. Submucosa, smooth muscles and striated muscles are less frequent [8]. Over 50% of cases is localized in the head and neck area, with the tongue being the most common site. Extremities are localization for 15% of cases. Other locations such as limbs, vulva, lung, breast, intestines, pituitary gland are also reported [3].

Abrikossoff tumor was initially described in 1926 by the Russian pathologist Alexei Ivanovich Abrikossofff. In 1854 Weber and Virchow described cluster of large cells with granular eosinophilic cytoplasm, but they believed it originated from skeletal muscle fibers, but Abrikossofff refuted myogenic origin examining the tumor in the tongue. The histological origin of GCT still remains uncertain. Different cell types have been postulated as histogenic of GCT including histiocytes, smooth muscle cells, fibroblasts, neural cells, neuroglia. Granular character of the tumor cells is related to accumulation of lysosomes in the cytoplasm in different fragmentation stages [8]. Recent studies show that neural/schwann — cell origin is the most accurate. Routine light microscopy is not sufficient to distinguish GCT from other tumors with granular cell morphology (e.g. basal cell carcinoma, melanoma, leiomyoma, leiomyosarcoma, dermatofibrosarcoma, angiosarcoma, fibrous histiocytoma and amyloblastoma). Expert pa-

thological assessment is essential, especially immunohistochemical staining, which is critical to identify its neural phenotype [8, 9]. The cytoplasmic granules are positive for periodic acid — Schiff (PAS) and for Sudan Black, but a key element is expression of S — 100 protein [2]. GCTs are also positive for neurospecific enolase, vimentine, CD56, CD57, CD63, CD68, epithelial membrane antigen (EMA), calretinin, protein gene product 9.5, nestin. Positivity for S-100, NSE, EMA and inhibin is concerned to be typical [3]. Our patient had characteristic histopathological presentation, expression of S-100 protein.

Tumors are not encapsulated and have imprecise borders. Neoplasm may infiltrate surrounding tissues — muscle, fat, subcutaneus, also nerves [10].

In our case there were no signs of local or remote metastases. The tumor grew slow for six months, without subjective symptoms.

GCTs usually display as a small (1–2 cm), painless, compact, nonulcerated, solitary nodule, but 5–25% of patients have multifocal lesions. They can occur synchronously or metachronously. Nodules grow slowly, usually with no alarming symptoms. Itching and pain were reported in tumors localized in hand and fingers [10]. It takes some time before patients start seeking help. Solitary lesions are more characteristic of children and adolescents [11]. Multiple GCT may be seen in the context of LEOPARD syndrome, phakomatoses, congenital heart defects, face anomalies. Bakos syndrome is a term proposed for cases of coexistence of multiply GCTs, with pulmonary valve stenosis, muscular hypotonia, joint hipermobility and mild facial dysmorphism.

Another entity related to granular cell pathology is congenital granular cell lesion (CGCL) also known as congenital granular cell epulis due to its gingival or alveolar localization. These lesions do not progress after birth. In immunohistochemical staining CGCL is negative for S — 100 [12].

Our patient had the size of the tumor only 5 mm but it may often find larger foci.

Macroscopic features of malignancy are: size (> 5 cm), ulceration, hemorrhage, necrosis, spindling, quick growth or acceleration of growth, local recurrence, metastases (local to distant lymphatic nodes and remote to lungs, brain, bones) [13]. In microscopic aspect: vesicular nuclei with large nucleoli, high mitotic index (more than 2 mitotic figures in 10 high power fields at 200 × magnification), perivascular infiltration, necrosis, apoptotic cells). High levels of proliferation marker Ki67 and expression of p53 protein may correlate with malignant course [8, 10]. Fatal cases with recurrence and remote metastases have been reported [14].

Patients die within 2–5 years after diagnosis, with approximately 40% mortality within 3 years [8]. That is why the accurate histological examination is mandatory.

The probability of recurrence depends on completeness of excision. Surgical excision with wide negative margin terminates the result of treatment. Risk of recurrence in margins free of tumor is 2–8% and about 20% when the margins are positive [4]. Methods with microscopic control of radicality and Mohs micrographic surgery (MMS) are preferred. In cases with malignant course treatment is a combination of surgery and chemotherapy [7].

CONCLUSION

Abrikossoff tumor in the extremity localization is uncommon neoplasm. Radical excision is the treatment of choice. It is possibility of recurrence and aggressive malignant course.

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