A Rare Neoplastic Growth on the Ear Lobe

Neobvyklá nádorová infiltrace ušního lalůčku

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Summary

We report a case of an 83-year-old previously healthy female patient presenting with a swiftly evolving erythematous violaceous, infiltrative, ulcerated onion-like mass with hyperkeratotic surface on the left ear lobe. The lesion was excised and resulted as an atypical fibroxanthoma, an extremely rare neoplastic growth, being a superficial variant of pleomorphic malignant fibrous histiocytoma. A brief review of diagnosis, treatment and prognosis is discussed.

Key words

skin – clear cell atypical fibroxanthoma – immunohistochemistry – differential diagnosis – head and neck neoplasms – neoplasms – prognosis

Souhrn

Prezentujeme případ 83leté ženy, dosud bez závažných onemocnění, u které byla zjištěna rychle rostoucí, zarudlá nafialovělá infiltrace levého ušního lalůčku, s hyperkeratotickým povrchem cibulovitého vzhledu a ojedinělou ulcerací. Po odstranění byla léze diagnostikována jako atypický fibroxanthom – zcela ojedinělý tumor představující kožní variantu maligního fibrózního histiocytomu. Článek v krátkosti popisuje diagnostiku, terapii a prognózu onemocnění.

Klíčová slova

kůže – světobuněčný atypický fibroxanthom – imunohistochemie – diferenciální diagnóza – nádory hlavy a krku – novotvary – prognóza

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Case report

An 83-year-old previously healthy female patient, agriculturist, presents with a history of an erythematous violaceous infiltrative, ulcerated onion-like mass with hyperkeratotic surface on the left ear lobe (Fig. 1–4). As the patient had a long history of chronic sun exposure and lived in one of the highest melanoma rates areas in the world [1], it was initially thought to be a metastatic melanoma by the surgeon.

The lesion was then completelly excised and sent for pathological analysis, with the result coming as a malignant ulcerated fusocellular neoplasia with negative margins. Further, an immuno-

histochemical analysis was performed and was negative for all markers, including protein S 100, all the cytokeratins, Melan A/MART 1, protein p53, CD 23 and desmin, compatible with an atypical fibroxanthoma, a very rare form of skin cancer. The atypical fibroxanthoma is a superficial variant of pleomorphic malignant fibrous histiocytoma [2]. Our case has followed the classic presentation as a head and neck tumor in an elderly individual, and to the best of our knowledge just one case in medical literature has been reported in a different topography-on the dorsum of the hand, described almost three decades ago [3].

In a retrospective analysis of Mohs surgery, only 0.2% of the malignant findings were fibroxantomas out of 42,279 patients [4].

Conclusion

Atypical fibroxanthoma normally appears as a swiftly growing nodular or nodulo-ulcerative lesion. It may be composed predominantly of either pleomorphic, spindle, epithelioid cells, or a mixture of these cells. The differential diagnosis includes pleomorphic dermal sarcoma, squamous cell carcinoma, malignant melanoma and leiomyosarcoma [5].

It occurs mostly in older adults and in sun exposed areas [6], with male pre-



Fig 1. A solitary erythematous nodule with hyperkeratotic and ulcerated surface on the left ear lobe (front view).



Fig. 2. A solitary erythematous nodule with prominent vessels, hyperkeratotic and ulcerated surface on the left ear lobe (side view).

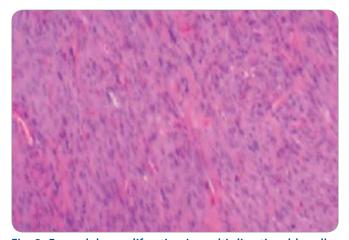


Fig. 3. Fusocelular proliferation in multi-directional bundles $(100\times)$.

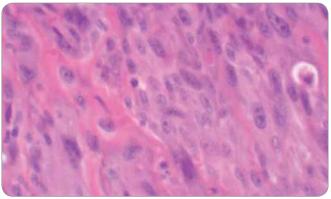


Fig. 4. Proliferation of elongated cells with poorly defined limits, dense eosinophilic cytoplasm, vesiculous or dense nuclei, irregular nuclear membrane and small to moderate diameter variation $(400\times)$.

dominance [7] and is a diagnosis of exclusion. The treatment is surgical and the preferred method is the aforementioned Mohs surgery [8]. Even though fibroxanthoma may be locally aggressive [9], the prognosis is usually very good if margins are adequate and these tumors rarely metastasise [7,10].

References

1. Naser N. Cutaneous melanoma: a 30-year-long epidemiological study conducted in a city in southern Brazil, from 1980–2009. An Bras Dermatol 2011; 86(5): 932–941.

- 2. Bedir R, Agirbas S, Sehitoglu I et al. Clear cell atypical fibroxantoma: a rare variant of atypical fibroxanthoma and review of the literature. J Clin Diagn Res 2014; 8(6): FD09–FD11. doi: 10.7860/JCDR/2014/8798.
- 3. Patterson JW, Konerding H, Kramer WM. "Clear cell" atypical fibroxanthoma. J Dermatol Surg Oncol 1987; 13(10): 1109–1114.
- **4.** Anderson HL, Joseph AK. A pilot feasibility study of a rare skin tumor database. Dermatol Surg 2007; 33(6): 693–696.
- **5.** Hussein MR. Atypical fibroxanthoma: new insights. Expert Rev Anticancer Ther 2014; 14(9): 1075–1088.
- **6.** Hammerschmidt M, Azevedo LM, Ruaro A et al. Case for diagnosis. Atypical fibroxanthoma. An Bras Dermatol 2012; 87(4): 647–648.
- 7. Wollina U, Schonlebe J, Ziemer M et al. Atypical fibroxanthoma: a series of 56 tumors and an unexplained uneven distribution of cases in southeast Germany. Head Neck. In press 2014. doi: 10.1002/hed.23673.
- 8. McCoppin HH, Christiansen D, Stasko T et al. Clinical spectrum of atypical fibroxanthoma and undifferentiated pleomorphic sarcoma in solid organ transplant recipients: a collective experience. Dermatol Surg 2012; 38(2): 230–239. doi: 10.1111/i.1524-4775.2011.02180.x
- **9.** Inskip M, Magee J, Weedon D et al. Atypical fibroxanthoma of the cheek-case report with dermatoscopy and dermatopathology. Dermatol Pract Concept 2014; 4(2): 77–80. doi: 10.5826/dpc.0402a16.
- **10.** Cooper JZ, Newman SR, Scott GA et al. Metastasizing atypical fibroxanthoma (cutaneous malignant histiocytoma): report of five cases. Dermatol Surg 2005; 31(2): 221–225.

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