

Primary cutaneous adenoid-cystic carcinoma of thigh found accidentally and presenting with the clinical picture of small pigmented (dark brown) cutaneous fibroma: A clinical case

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CASE STUDY

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ABSTRACT

The primary cutaneous adenoid-cystic carcinoma represents a very rare neoplasm - less than 100 similar cases are reported in the literature until now. It affects persons of middle age or elder the female is involved a little more often the local relapses are observed in about half of the cases and it metastasizes most frequently in the regional lymph nodes and lungs.

We present a case of a 31-year-old woman with primary adenoid-cystic carcinoma of skin which was treated for a

pigmented cutaneous fibroma. This required reoperating the patient.

The clinical manifestation of primary cutaneous adenoid-cystic carcinoma may imitate benign pigmented cutaneous fibroma. The final diagnosis of this disease is made by a pathologist based on the pathomorphological exam and immunohistochemistry. The basic method for treatment in these cases is the extensive local excision in a radius of 2cm from the lesion to avoid the risk of local relapse.

Key Words

Primary cutaneous adenoid-cystic carcinoma, cutaneous fibroma, skin cancer

Implications for Practice:

1. What is known about this subject?

The Primary Cutaneous Adenoid-Cystic Carcinoma is a very rare malignant tumour.

2. What new information is offered in this case study?

We present a case of a 31-year-old woman with primary adenoid-cystic carcinoma of skin which looks like small pigmented cutaneous fibroma.

3. What are the implications for research, policy, or practice?

The adenoid-cystic carcinoma very rare can originates from the eccrine sweat glands of the skin.

Background

The primary cutaneous adenoid-cystic carcinoma represents a very rare neoplasm.^{1,2} It originates from the eccrine sweat glands of the skin, and at the same time, it has slow and progressive growth. It represents as a hard, not well-defined skin lesion. The most frequent location is the scalp. Persons of middle age are predominantly affected and they are most frequently of females.^{1,3,4} The local relapses are observed in about half of the cases^{1,3} and it metastasizes most frequently in the regional lymph nodes and lung.^{3,5-12} Surgical excision is the basic method of treatment of that carcinoma, while radiotherapy is applied for prevention of subsequent local relapses.¹²

We present a case of a 31-year-old woman with primary adenoid-cystic carcinoma of skin which was treated for a pigmented cutaneous fibroma. This required reoperating the patient.

Case details

We present a 31-year-old woman with comorbidities of arterial hypertension and chronic bronchitis, who palpated a hard, but not a painful lump, in the skin of upper inner part of her right thigh - with sizes of a pea - several months ago. Its colour was dark brown. The patient submitted to an examination by a dermatologist on that occasion - the doctor made a diagnosis of pigmented cutaneous fibroma and directed her for its surgical excision. The patient was admitted at the Clinical Department of Surgery for performing the operation. Nothing abnormal was detected by the laboratory examinations the patient underwent. Chronic bronchitis was found as evident from the radiography of the lung. A hard, intradermal, dark brown, well-defined tumour formation was found from the physical examination, with sizes of 1/1cm, localized in the skin of medial upper one-third of the right thigh. That formation was assessed preoperatively as pigmented, intradermal, cutaneous fibroma and was removed surgically together with 2 mm of the healthy, surrounding skin. The preparation was sent for examination by specialists of histologic pathology with the following result: Material from the skin and subcutaneous tissue with infiltration in the deep derma by strands and nests of basaloid cells with glandular, cystic, and cribriform structure amidst fibro-vascular stroma, with no connection to the epidermis. Periodic acid-Schiff stain positive (PAS+) and alcian blue stain positive (Alcian+) matters were found in the glandular and cystic spaces. The tumour cells were positive for p63, and negative for CK7, CD117, and CEA. Based on the morphology and immunohistochemistry, it was assumed that this is a case of

primary cutaneous adenoid cystic carcinoma (Figure 1).

That required performing of reexcision at the site of the primary tumour in a radius of 2cm from the scar of the previous operative intervention with a view to the great risk of local relapse on that type of carcinoma. Based on the examination of the flap from reexcision that was carried out by specialists of histologic pathology, there were no data found of residual tumour. The patient was referred to an oncological dermatologist for follow-up.

Discussion

The adenoid-cystic carcinoma most commonly affects the major and minor salivary glands, but it may be found in the lacrimal glands, external auditory canal, uterus, cervix, vulva, respiratory tract, thymus, breast, prostate, oesophagus, and skin.^{13,14} The primary cutaneous adenoid-cystic carcinoma is a very rare tumour. It was described for the first time by Boggio in the year 1975.^{1,15} Less than 100 similar cases are reported in the literature until now.^{1,2} The origin of this neoplasm was determined to be the eccrine glands of the skin. Its size varies from 0.5–8.0cm, while in 41 per cent of all cases it involved the scalp.^{1,3} Other frequently found localizations are the abdomen, thoracic cage, back, eyelids, and perineum. It presented clinically as a hard, non-pigmented skin node with no distinct borders and of slow growth, localized intra- or subdermally, which most commonly manifests no symptoms, however, it may be accompanied by secondary alopecia, tenderness, or itching. This neoplasm affects persons of middle age or elder and the average age to diagnose the disease is 59 years. The female is involved a little more often (54 per cent).^{1,3,6} Local relapse developed within 58 months after the operative treatment in 44 per cent of the cases.^{1,3} Metastasis of the primary cutaneous adenoid-cystic carcinoma is a very rare, but when it happens lung and regional lymph nodes are most often affected (in 20 per cent of the cases).^{3,5-12} For the accomplishment of diagnosis of that disease, the clinical picture is definitely not sufficient - the result reached by specialists of histologic pathology after biopsy or excision is crucial as well.¹ A basic aspect in the treatment of that carcinoma is the extensive surgical excision of the tumor within non-affected tissues, usually in a radius of 2cm, while it if needed - may be followed by local radiotherapy.¹²

Some interest in our case arouses the fact that patient is 31-year-old, which is quite under the average age for development of that disease (59 years of age).^{1,3,6} Based on the detailed study of world literature for the primary cutaneous adenoid-cystic carcinoma on this matter, we found only one case - similar to ours - with involvement of

thigh, and at that of the left one.¹ Another very interesting aspect is that in this case, the carcinoma had its course with the clinical picture of the pigmented cutaneous fibroma (a hard, intradermal, dark brown, well-defined and with even borders tumour formation, with sizes of 1/1cm) and that it by no means was preliminarily suspected. That was the reason to perform a reexcision of the scar from the previous excision in a radius of 2cm, after which we learned about the result of the primarily removed tissue examination carried out by specialists of histologic pathology.

Conclusion

In conclusion, the primary cutaneous adenoid-cystic carcinoma is a very rare neoplasm. The clinical manifestation of this type of carcinoma may imitate benign pigmented cutaneous fibroma. The final diagnosis of this disease is made by pathologist based on the pathomorphological exam and immunohistochemistry. The basic method for treatment in these cases is the extensive local excision in radius of 2cm from the lesion to avoid the risk of local relapse.

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PEER REVIEW

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CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

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PATIENT CONSENT

The authors, *Strashilov S, Slavchev S, Nanev V, Ivanova D, Ivanov M, Ivanov I, Yordanov A*, declare that:

1. They have obtained written, informed consent for the publication of the details relating to the patient(s) in this report.
2. All possible steps have been taken to safeguard the identity of the patient(s).
3. This submission is compliant with the requirements of local research ethics committees.

Figure 1: Histology of primary cutaneous adenoid-cystic carcinoma

