CASE REPORT

MRI of a microcystic adnexal carcinoma of the skin mimicking a fibrous tumour: case report and literature review

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ABSTRACT. Microcystic adnexal carcinoma of the skin is a very rare malignant tumour arising from the sweat glands. As far as we know, the MRI features of this tumour have not been described in the literature before. In this report we present the MRI features and pathological description of a case of a microcystic adnexal carcinoma in the cheek that was incidentally imaged during brain MRI examination. A review of the relevant literature as well as a discussion of MRI of skin tumours is also presented.

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Microcystic adnexal carcinoma is a very rare skin tumour, with only approximately 300 cases reported in the literature [1]. It is generally believed to arise from the eccrine sweat glands and has a definite predilection to the head and neck region. Owing to its rarity and its benign clinical appearance, the diagnosis is usually delayed and the final diagnosis is pathological examination rather than clinical [2].

The radiological findings of most skin tumours, even the far more common entities, are not well documented in the literature. As far as we know, the MRI findings of microcystic adnexal carcinoma have not been described before. In this report we present the MRI findings and pathological description of a case of microcystic adnexal carcinoma of the skin of the cheek.

Case report

A 67-year-old male complaining of recent tinnitus and vertigo was referred for MRI to rule out the presence of a central or cerebellopontine angle (CPA) lesion. Imaging was performed with a 1.5 Tesla MRI (MAGNETOM, Espree, Siemens Medical Solutions, Erlangen, Germany) using the following sequences. Axial T_2 weighted turbo spin-echo sequence: repetition time (TR), 5510 ms; echo time (TE), 105 ms; slice thickness (ST), 5 mm; slice spacing, 5.5 mm. Axial T_1 weighted three-dimensional (3D) MP-RAGE (magnetisation-prepared rapid acquisition gradient echo) sequence: TR, 2250 ms; TE, 2.94 ms; ST, 1.2 mm. Axial T_1 weighted 3D MP-RAGE sequence with fat suppression after intravenous injection of contrast (Magnivest, Bayer Schering Pharma, Berlin, Germany) in

a dose of 0.1 mmol kg⁻¹ body weight. Coronal contrast-enhanced T_1 weighted spin echo sequence; TR: 628 ms, TE: 8.8 ms, ST: 5 mm slice spacing=5.5 mm.

There was no abnormality detected in the brain or CPA but an incidentally imaged lesion was visible in the right cheek. The lesion involved mainly the subcutaneous region with a wide base inseparable from the overlying skin. The skin line itself appeared intact, especially on T_2 weighted images. The lesion stood out clearly against the surrounding subcutaneous fat but its borders were partially ill defined. No invasion of the deep tissues (muscle or bone) was detected. It measured 2.7 × 2 × 1.5 cm. The signal intensity was generally homogeneous. On T_1 weighted images, the lesion was hypo-intense compared with fat. On T_2 weighted images it was also of low-signal intensity, iso- to hypo-intense to the muscles (Figure 1). There was no evidence of areas of necrosis, haemorrhage or calcification. After contrast administration it showed faint homogeneous enhancement (Figures 2 and 3). Surrounding subcutaneous fat appeared normal. These MRI findings were suggestive of an indeterminate lesion and malignancy could not be excluded. A surgical biopsy was advised.

1 month after the MRI examination, surgical excision of the lesion was done and pathological examination revealed a malignant tumour with infiltration of the surgical margins. The surgical specimen included the skin and subcutaneous tissues. Microscopically the tumour consisted of solid to cystic structures infiltrating a relatively hypocellular fibrotic stroma of the dermis and subcutis (Figure 4). Cytologically the tumour cells revealed relatively bland nuclei and only few mitoses. The infiltration of a perineural sheath was also seen. No basal retraction or peripheral nuclear palisading was detected, nor ulceration of the covering epidermis with regular layering and keratosis. The diagnosis was microcystic adnexal carcinoma.

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Figure 1. Axial turbo spin-echo T_2 weighted image shows the lesion in the subcutaneous tissue of the right cheek, with wide base inseparable from overlying skin. The lesion is of homogeneous low-signal intensity, hypo- to isointense to muscles. Note also the intact overlying skin line (arrow).

Discussion

Imaging plays a small role in pre-operative diagnosis of skin tumours and is usually reserved for metastatic work-up of already diagnosed skin malignancies. This is in contrast to many other organs of the human body and the reason is simple: skin tumours are readily accessible to clinical examination and surgical biopsy. MRI, however, may have great potential as an auxiliary diagnostic tool for skin tumours because it is non-invasive, safe and has excellent repeatability. Characterisation of the nature of a skin tumour with MRI and definitive exclusion of malignancy is not always possible, but confirmation of some benign entities such as an epidermal

Figure 2. Axial magnetisation-prepared rapid acquisition gradient echo contrast-enhanced T_1 weighted image with fat suppression shows mild enhancement of the lesion (arrows).

Figure 3. Coronal spin-echo contrast-enhanced T_1 weighted image shows the mildly enhancing, homogeneous lesion, well demarcated from the surrounding high-signal intensity subcutaneous fat (arrows).

cyst, subcutaneous lipoma or a vascular malformation can change the clinical decision. MRI is currently used for evaluation of the extent and deep invasion in some skin tumours. Other less common indications include evaluation of suspected recurrence or detection of a malignant change in a chronic skin lesion such as Marjolin's ulcer. With ongoing technical progresses in MRI improving the resolution, it becomes more and more suitable for imaging of skin tumours. Moreover, the continuous rise in the number of MRI examinations for variable indications increases the possibility of incidental imaging of skin tumours.

Skin tumours comprise a vast array of benign, pre-malignant and malignant lesions as well as cutaneous metastases. Primary tumours are classified into melanomatous

Figure 4. High-power photomicrograph (magnification $\times 40$) of the tumour shows a microcystic pattern of growth, containing trichilemmal-type keratin. The nuclei of the tumour cells lack nucleoli and exhibit some increased mitotic activity.

and non-melanomatous. Non-melanomatous tumours can be further classified on the basis of their origin as epidermal, dermal and adnexal. The commonest malignant non-melanomatous tumours are the basal and squamous cell carcinomas [3]. Skin adnexal tumours are characterised by differentiation into one of the adnexal components, namely the pilosebaceous unit, apocrine and eccrine sweat glands. Some tumours show differentiation into more than one cell line [2]. Most sweat gland tumours are uncommon, with some types considered very rare [3]. Their further classification is complex and the terminology used is variable owing to their unclear pathogenesis, so that one tumour may be described with many different terms. Coexistence of eccrine and apocrine differentiation in one tumour adds more difficulty in description and terminology [2]. Microcystic adnexal carcinoma was first described as a distinct entity by Goldstein et al in 1982 [4]. It is usually classified under tumours of eccrine sweat gland origin, although the definite cell of origin is thought to be a pluripotent adnexal keratinocyte capable of differentiation into both sweat gland and hair follicle lines [2]. It tends to affect the central face of middle-aged adults commonly presenting as an asymptomatic firm, non-ulcerated nodule with apparent stable size over long periods. It is, however, locally aggressive, typically extending beyond the clinically visible margin, with deep subcutaneous extension and characteristically perineural invasion. Lymph node or distant metastases are exceedingly unlikely [2].

Reviewing the literature, we did not find any descriptions of the imaging features of microcystic adnexal carcinoma, but we did find sporadic case reports describing MRI features of other tumours of the eccrine sweat gland origin. The MRI findings in three cases of syringoma were published in separate case reports and consisted of a subcutaneous [5, 6], or subfascial [7], well-circumscribed mass, with intermediate signal intensity on proton density [5], or low signal intensity on T_1 weighted images [6, 7]. All three lesions showed high signal intensity on T_2 weighted and STIR (short-tau inversion recovery) images with a slight heterogeneous appearance. After contrast administration there was diffuse enhancement [6, 7]. Porocarcinoma was described on MRI as a diffuse subcutaneous mass with ill-defined infiltrative borders, low signal intensity on T_1 weighted images, inhomogeneous intermediate-to-high signal intensity on T_2 weighted images and with high inhomogeneous contrast enhancement [8]. Hidradenoma was described on MRI as a partially cystic mass with occasional haemorrhage and a mural nodule with or without enhancement [9], another reported case appeared as a solid diffusely enhancing mass with low signal intensity on T_1 and high signal intensity on T_2 weighted images, respectively [10]. A case of multiple eccrine spiradenoma was described on MRI as multiple separate and confluent dermal and subcutaneous nodules of homogeneous low signal intensity on T_1 weighted images and high signal intensity on STIR images [11].

The MRI findings in this case are different because the tumour is of homogeneous low signal intensity on both T_1 and T_2 weighted images. Finding a lesion with signal intensity similar or lower than that of muscle on T_2 weighted images is usually regarded as more specific, and is supposed to limit the differential diagnosis. For example, low signal intensity in a soft-tissue tumour is

considered to indicate the presence of a significant fibrous component, calcification or haemosiderin content [12]. The low signal intensity of fibrous tissue is explained by the relative lack of mobile protons associated with the hypocellular densely collagenous matrix [12]. Correspondingly, the differential diagnosis of such a tumour as in this case based on signal intensity and after exclusion of scar tissue will include only fibrous tumours (examples are fibroma, fibrosarcoma, desmoids and nodular fasciitis).

But is low T_2 signal intensity alone enough for diagnosis of a fibrous tissue tumour? We reviewed previous studies discussing T_2 signal intensity of some fibrous tumours with histopathological correlation. One example is the relatively uncommon solitary fibrous tumour of the head and neck region where, not surprisingly, most of the reported cases showed low T_2 signal intensity [13]. In a few other cases, however, a rather uniform high T_2 signal intensity was reported, and was attributed to the variation in cellularity and amount of collagen [13, 14]. Another example for the T_2 signal intensity of fibrous tissue tumours are two cases of aggressive fibromatosis studied by the same group and showing opposite signal intensities on T_2 weighted images; histologically they contained the same collagen amount but the difference was in cellularity with high and low T_2 signal intensities seen in hyper- and hypocellular tumours, respectively [15]. On the other hand, it is worth noting that low T_2 signal intensity was also observed in high-grade malignant soft-tissue tumours, and this was attributed to their high cellularity and increased nucleocytoplasmic ratio, both causing decreased free intra- and extracellular water [16]. From above it appears that a low T_2 signal intensity tumour is not always a fibrous tumour and high T_2 signal intensity does not exclude a fibrous tumour. In our case, the low signal intensity on T_2 weighted images can similarly be explained as a result of the high amount of hypocellular fibrohyalinised stroma in between neoplastic structures. We believe, however, that a radiological-pathological correlation over large series of tumours is needed to explain these relations between cellularity, cytoplasm-nuclear ratio, collagen and fibrous tissue in tumour stroma on one hand and T_2 signal intensity on the other. Until then, T_2 signal intensity should be considered to vary according to variation in proportions of different histological components and not according to specific tumour histological type.

While the World Health Organization (WHO) classification and most of the radiology literature do not include any of the skin tumours in the classification or differential diagnosis of soft-tissue tumours [17], other authors do include some skin tumours (dermal and appendageal) together with soft-tissue tumours (from subcutaneous tissue) in one differential diagnosis [18]. This seems logical when discussing superficial soft-tissue masses, especially from a radiological point of view, where location rather than actual origin is usually appreciated. However, it may cause confusion between terms without a clear definition of superficial soft-tissue region. It may also imply the application of some rules adopted for soft-tissue imaging that may not directly apply for skin tumours. For example, the size and depth of a soft-tissue tumour, greater than 5 cm, deep location

or crossing of the fascia are considered as predictors of malignancy; whereas a superficial tumour less than 3 cm that does not cross the fascia is more likely benign [19]. Whether true or not, these and other guidelines and rules for soft-tissue imaging clearly do not fit the imaging of skin tumours. It seems more logical to separate the differential diagnosis of skin tumours, most of which lack well-documented MRI findings, from the relatively well-studied soft-tissue tumours. An attempt for separate differential diagnosis of skin tumours based on their MRI appearances was described by Kim et al [20]. Other studies are definitely needed to assess first the ability of MRI to identify the actual origin of tumours whether cutaneous (epidermal, dermal or adnexal) or subcutaneous soft-tissue tumours extending to the skin surface, and second how would the MRI appearance help in the characterisation of skin tumours.

Conclusion

MRI appears to be well suited for the imaging of skin tumours, and its role is expected to rise in the near future. Radiologists should be aware of the differential diagnosis and radiological appearances of skin tumours, which should be studied and classified separately from soft-tissue tumours. MRI shows the exact size of the lesion, its exact location and the deep extent of the lesion, and can exclude some benign lesions with certainty. Any skin lesion that appears non-specific or indeterminate on MRI should be further evaluated either by biopsy or follow-up.

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References

1. Yu JB, Blitzblau RC, Patel SC, Decker RH, Wilson LD. Surveillance, epidemiology, and end results (SEER) database analysis of microcystic adnexal carcinoma (sclerosing sweat duct carcinoma) of the skin. *Am J Clin Oncol* 2010;33:125–7.
2. Obaidat NA, Alsaad KO, Ghazarian D. Skin adnexal neoplasms-part 2: an approach to tumours of cutaneous sweat glands. *J Clin Pathol* 2007;60:145–59.
3. McGuire JF, Ge NN, Dyson S. Nonmelanoma skin cancer of the head and neck I: histopathology and clinical behaviour. *Am J Otolaryngol* 2009;30:121–33.
4. Goldstein DJ, Barr RJ, Santa Cruz DJ. Microcystic adnexal carcinoma: a distinct clinicopathologic entity. *Cancer* 1982; 50:566–72.
5. Nicolaou S, Dubec JJ, Munk PL, O'Connell JX, Lee MJ. Malignant chondroid syringoma of the skin: magnetic resonance imaging features. *Australas Radiol* 2001;45:240–3.
6. Kerimoglu U, Aydingoz U, Ozkaya O, Aksu AE, Ergen FB. MRI of a benign chondroid syringoma. *Br J Radiol* 2006;79: 59–61.
7. Adachi T, Oda Y, Sakamoto A, Saito T, Tamiya S, Hachitanda Y, et al. Mixed tumour of deep soft tissue. *Pathol Int* 2003;53:35–9.
8. Iannicelli E, Galluzzo A, Salvi PF, Ziparo V, David V. A large porocarcinoma of perineal region: MR findings and review of the literature. *Abdom Imaging* 2008;33:744–7.
9. Mullaney PJ, Becker E, Graham B, Ghazarian D, Riddell RH, Salonen DC. Benign hidradenoma: magnetic resonance and ultrasound features of two cases. *Skeletal Radiol* 2007;36:1185–90.
10. Reier AR, Farooki S, Ashman CJ, Miles L. MR imaging appearance of plantar eccrine acrospiroma (sweat gland tumour). *AJR Am J Roentgenol* 2002;179:1079–80.
11. Han YD, Huan Y, Deng JL, Zhang YG, Zhang CH. MRI appearance of multiple eccrine spiradenoma. *Br J Radiol* 2007;80:27–9.
12. Wu JS, Hochman MG. Soft-tissue tumours and tumour like lesions: a systematic imaging approach. *Radiology* 2009; 253:297–316.
13. Ganly I, Patel SG, Stambuk HE, Coleman M, Ghossein R, Carlson D, et al. Solitary fibrous tumours of the head and neck: a clinicopathologic and radiologic review. *Arch Otolaryngol Head Neck Surg* 2006;132:517–25.
14. Shin JH, Sung IY, Suh JH, Yang SO, Jeong YK, Lee JH, et al. Solitary fibrous tumour in the buccal space: MR findings with pathologic correlation. *AJNR Am J Neuroradiol* 2001; 22:1890–2.
15. Sundaram M, McGuire MH, Schajowicz F. Soft-tissue masses: histologic basis for decreased signal (short T_2) on T_2 -weighted MR images. *AJR Am J Roentgenol* 1987;148: 1247–50.
16. De Schepper AM, De Beuckeleer L, Vandevenne J, Somville J. Magnetic resonance imaging of soft tissue tumours. *Eur Radiol* 2000;10:213–23.
17. Vilanova JC, Woertler K, Narváez JA, Barceló J, Martínez SJ, Villalón M, et al. Soft-tissue tumours update: MR imaging features according to the WHO classification. *Eur Radiol* 2007;17:125–38.
18. Beaman FD, Kransdorf MJ, Andrews TR, Murphey MD, Arcara LK, Keeling JH. Superficial soft-tissue masses: analysis, diagnosis, and differential considerations. *Radiographics* 2007;27:509–23.
19. Galant J, Martí-Bonmatí L, Soler R, Saez F, Lafuente J, Bonmatí C, et al. Grading of subcutaneous soft tissue tumours by means of their relationship with the superficial fascia on MR imaging. *Skeletal Radiol* 1998;27:657–63.
20. Kim JH, Kim JY, Chun KA, Jee WH, Sung MS. MR imaging manifestations of skin tumours. *Eur Radiol* 2008;18:2652–61.