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CASE REPORT

Surface-type chondromyxoid fibroma in an elderly patient: a case report and literature review

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Abstract

Chondromyxoid fibroma (CMF) is a rare benign bone neoplasm that typically occurs in young adults. Juxtacortical or surface-type CMF are rarer still and we present the case of a surface-type CMF in a 78-year-old woman, with only one other case described in a patient of a similar age previously. This patient was an otherwise healthy woman who presented for evaluation of a palpable lump in the anterior proximal tibia. Initial radiographs obtained demonstrated a focal soft tissue fullness immediately anterior to the anterior cortex of the proximal tibia, which contained faint chondroid-like matrix internally. There was associated scalloping of the anterior tibial cortex. MRI confirmed the presence of a juxtacortical, enhancing lesion. Subsequent excisional biopsy was performed and histopathology demonstrated features, which was consistent with surface-type CMF. At a 6-month follow-up the patient remained free of recurrence. In a patient of this age, paraosteal chondrosarcoma should be excluded. Surface-type CMF, although rare, has been described in older patients and while it is unlikely to feature in a list of differential considerations on initial imaging, awareness of the entity is important.

Keywords Juxtacortical · Surface type · Chondromyxoid fibroma

Introduction

Chondromyxoid fibroma is a rare and benign bone neoplasm that typically occurs in young adults, with the majority of cases occurring in the second and third decades of life. CMF accounts for less than 1% of all primary bone tumors, and juxtacortical, or surface, types are only reported in a small number of case reports and case series within the literature [1–18]. Conventional CMF is typically intramedullary and metaphyseal in location with a predilection for the long tubular bones of the lower extremities [19]. The most common locations include the proximal tibia, distal femur, pelvic bones, and small bones of the hands and feet occurs. Radiographically, the lesion is usually eccentrically located, lucent, and expansile with a thin, well-circumscribed sclerotic rim [19, 20]. We encountered a CMF that occurred on the cortical surface of an elderly female patient. The possibility of CMF was not initially raised on imaging due to the patient age and the juxtacortical location of the lesion. This case report reviews the imaging findings of surface-type CMF and reviews the previously reported cases in the literature.

Case report

An otherwise healthy 78-year-old woman presented for evaluation of right anterior knee mass. The mass had been present for approximately 5 years and she denied any history of trauma to the region.

The patient reported some mild tenderness on direct palpation of the mass but was otherwise asymptomatic. She presented for further assessment of the mass as she felt it had slightly increased in size. Clinical examination revealed a palpable firm mass at the level of the tibial tuberosity with minimal tenderness elicited on palpation. There was no discoloration or ulceration of the overlying skin. Range of motion at the knee joint was within normal limits. Radiographs obtained revealed a soft tissue mass anterior to the tibial tuberosity with saucerization of the underlying bone. There were faint regions of mineralization within the lesion (Fig. 1). An MRI was

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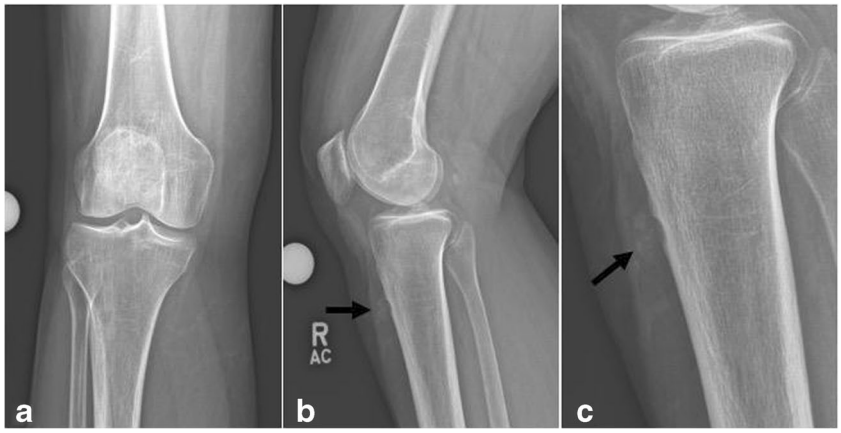

Fig. 1 **a**, **b**, **c** Frontal, lateral, and magnified lateral views of the right knee demonstrate a well-circumscribed lytic lesion on AP view, with an extraosseous soft tissue component in the anterior tibial soft tissues on lateral view. There is saucerization of the underlying cortical bone and faint chondroid-like matrix within the lesion (*arrow*)

performed and showed a well-circumscribed rounded lesion abutting the anterior tibial cortex just inferior to the tibial tuberosity. The lesion was T2 hyperintense and T1 isointense to muscle with diffuse enhancement on post contrast images (Figs. 2 and 3). There was no associated periosteal reaction, perilesional bone marrow, or soft tissue edema. As also seen on radiograph, there was smooth scalloping of the underlying cortex, and the lesion did not extend into the medulla. At the time of imaging, a juxtacortical chondroma or fibrous tumor was thought likely; however, given the patient's older age, the possibility of a low-grade chondrosarcoma was raised.

An excisional biopsy was performed and the pathology demonstrated a benign tumor composed of stellate and spindle-shaped cells associated with a myxoid matrix that was pseudo-lobular and demonstrated cellular condensation at the periphery of the pseudo-lobules (Figs. 4 and 5). Matrix calcifications were present with additional foci of reactive bone (Fig. 6). Immunohistochemistry was strongly positive for vimentin (Fig. 7), weakly positive for smooth muscle actin, and negative for S100 and beta-catenin. The histomorphologic findings combined with radiologic findings were diagnostic of a surface-type chondromyxoid fibroma.

Discussion

Chondromyxoid fibroma was first descried by Jaffe and Lichtenstein as a cartilage-forming lesion with myxoid and fibrous components, with the authors noting at the time how this tumor can be easily confused with a chondrosarcoma [4]. These tumors typically form at the metaphysis close to the growth plate and may extend to the epiphyseal line. They are slightly more common in males and are seen most frequently in the second or third decades. In a case series of 278 CMF, a large range in ages was seen with patients ranging from 6 to 87 years of age and the average age was 31 [19]. These tumors tend to be expansile, with thin, well-circumscribed sclerotic margins. Periosteal reaction or cortical breakthrough may rarely occur. Matrix calcification can be

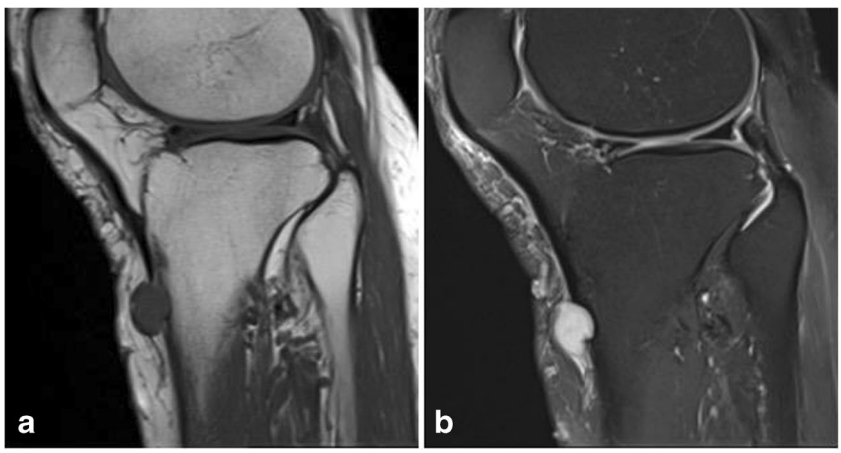

Fig. 2 **a** Sagittal views of the knee on T1-weighted imaging. The lesion is isointense to muscle. **b** Sagittal views of the knee on fat-saturated T2-weighted imaging. The lesion is homogenously hyperintense with preservation of the inner cortex. No intra-medullary extension, periosteal reaction, or bone marrow edema were observed

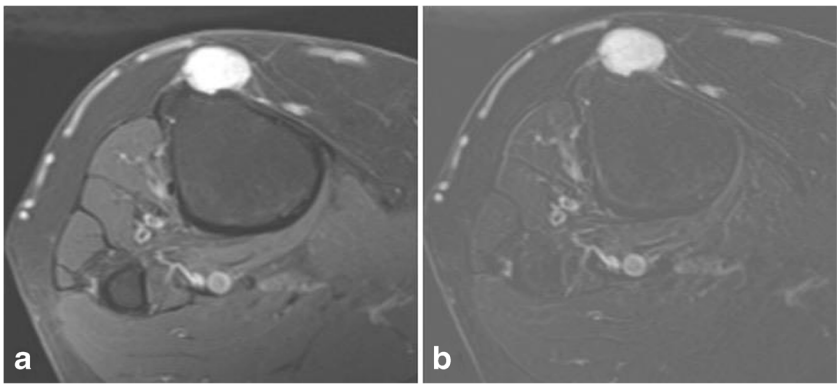

Fig. 3 **a**, **b** Axial fat-saturated T1-weighted imaging and axial fat-saturated T1-weighted imaging with subtractions demonstrate diffuse homogenous enhancement of the lesion

seen and has been described in between 2 and 16% of cases [21]. Typically, these tumors are eccentrically intramedullary but a number of case reports and case series describe lesions that arise from the surface of the bone. Surface-type or juxtacortical chondromyxoid fibromas refer to tumors that arise from within the cortex (intracortical), under the

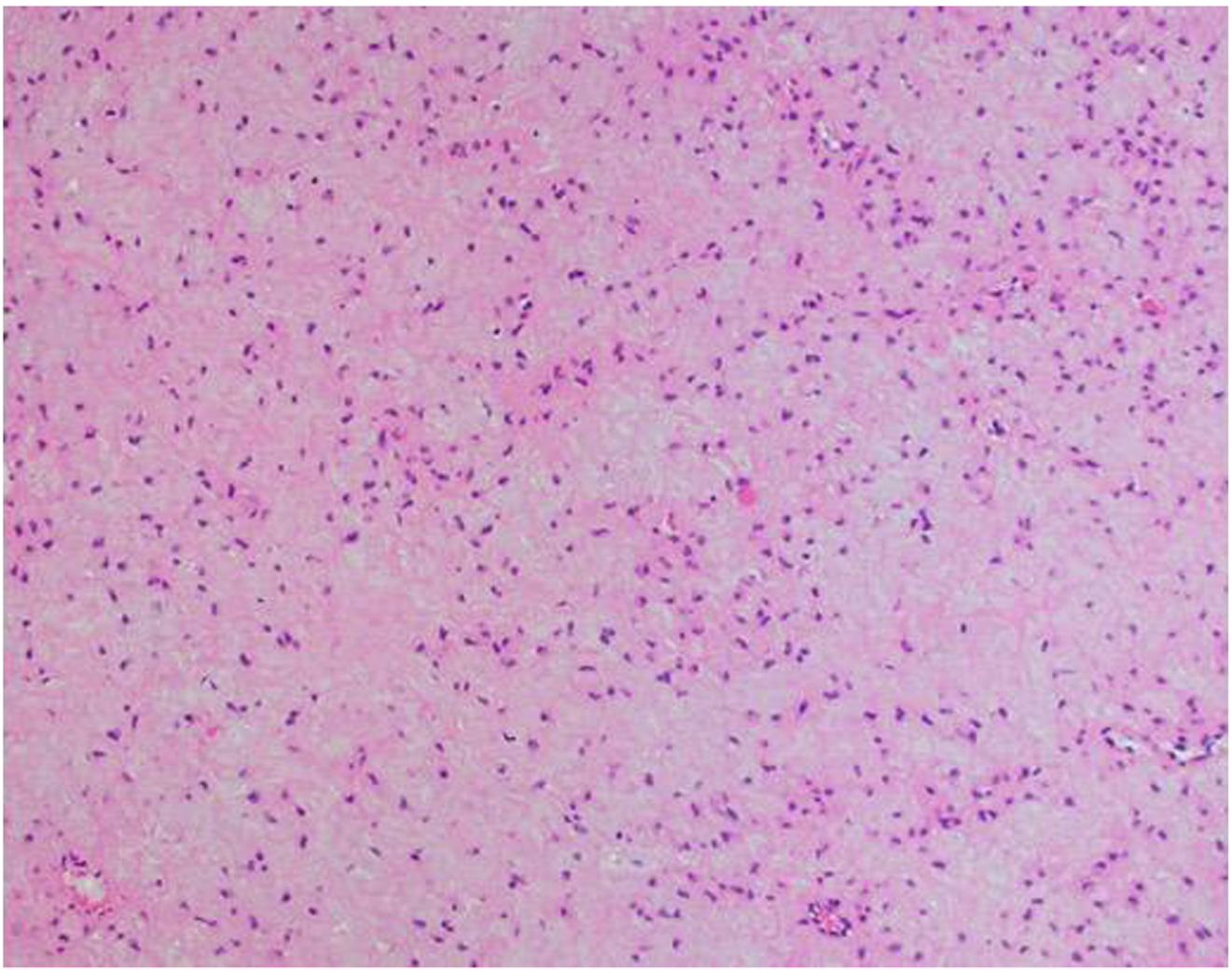

Fig. 4 Low-power magnification demonstrates a myxoid (*faint bluish stain*) spindle cell lesion with variable cellularity

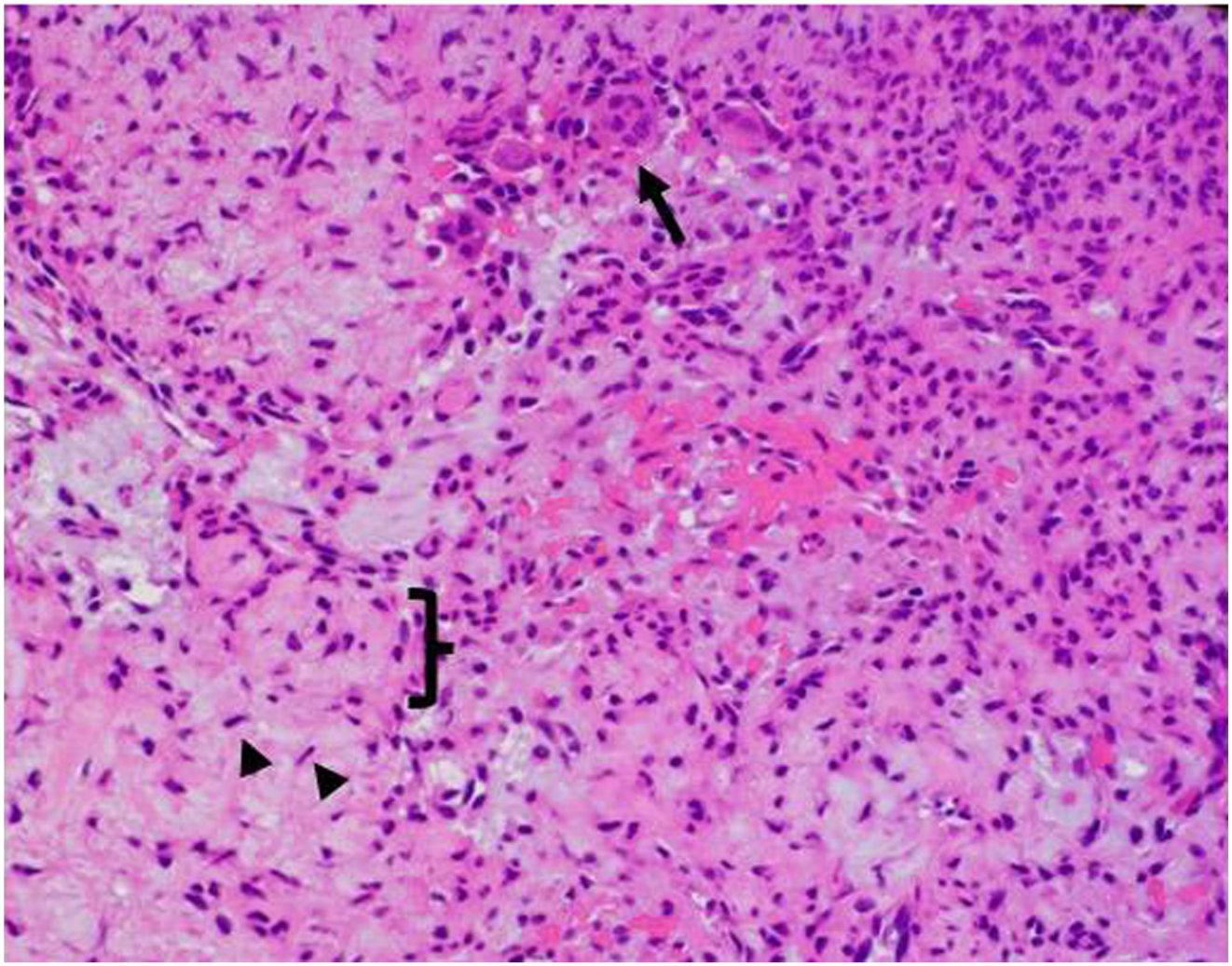

Fig. 5 High-power magnification demonstrates stellate and spindle-shaped cells with pseudo-lobules and increased cellularity at the periphery of the pseudo-lobules (*bracket*). Periphery of the lesion showing characteristic giant cell (*arrow*) present amongst hypercellular ovoid to spindle cells. Scattered stellate-shaped elongated nuclei present (*arrowheads*)

periosteum (subperiosteal), within the periosteum (periosteal) or fibroblastic tissues along the outer periosteum (paraosteal).

A literature review was performed on all previously reported juxtacortical or surface-type chondromyxoid fibromas. Baker et al. published the largest case series describing 20 cases collected over a 47-year period and suggested that surface-type CMF had some differentiating features to conventional CMF. Only two patients in this case series had their imaging findings described however. Firstly, the presence of calcification was found more frequently within surface-type CMF than in the typical intramedullary lesions. Secondly, the median age was higher within this cohort of patients at 40 years (average age was also 40).

However, when the ages from other published case series and case reports were combined, the average age drops to 36 years and the median age is 32 years. This may be accounted for by the wide range in patient age from 5 to 81 years across the literature. In addition, case reports of these tumors are also more likely to be reported when presenting unusually in patients at the extremes of ages, resulting in a selection-type bias. This reported case is the second eldest patient in which this tumor has been described. In all other case reports, 15 out of 40 cases (41%) of the patients were over the age of 40, with over just over half of these occurring in the 6th decade of life or more.

Of the reported cases, 61% of the cases occurred in male individuals. However, women had a higher average age of 41 years compared to the male average age of 33 years. Less than half (44%) of the surface-type CMFs occurred in the proximal tibia. Interestingly, when a surface-type CMF is

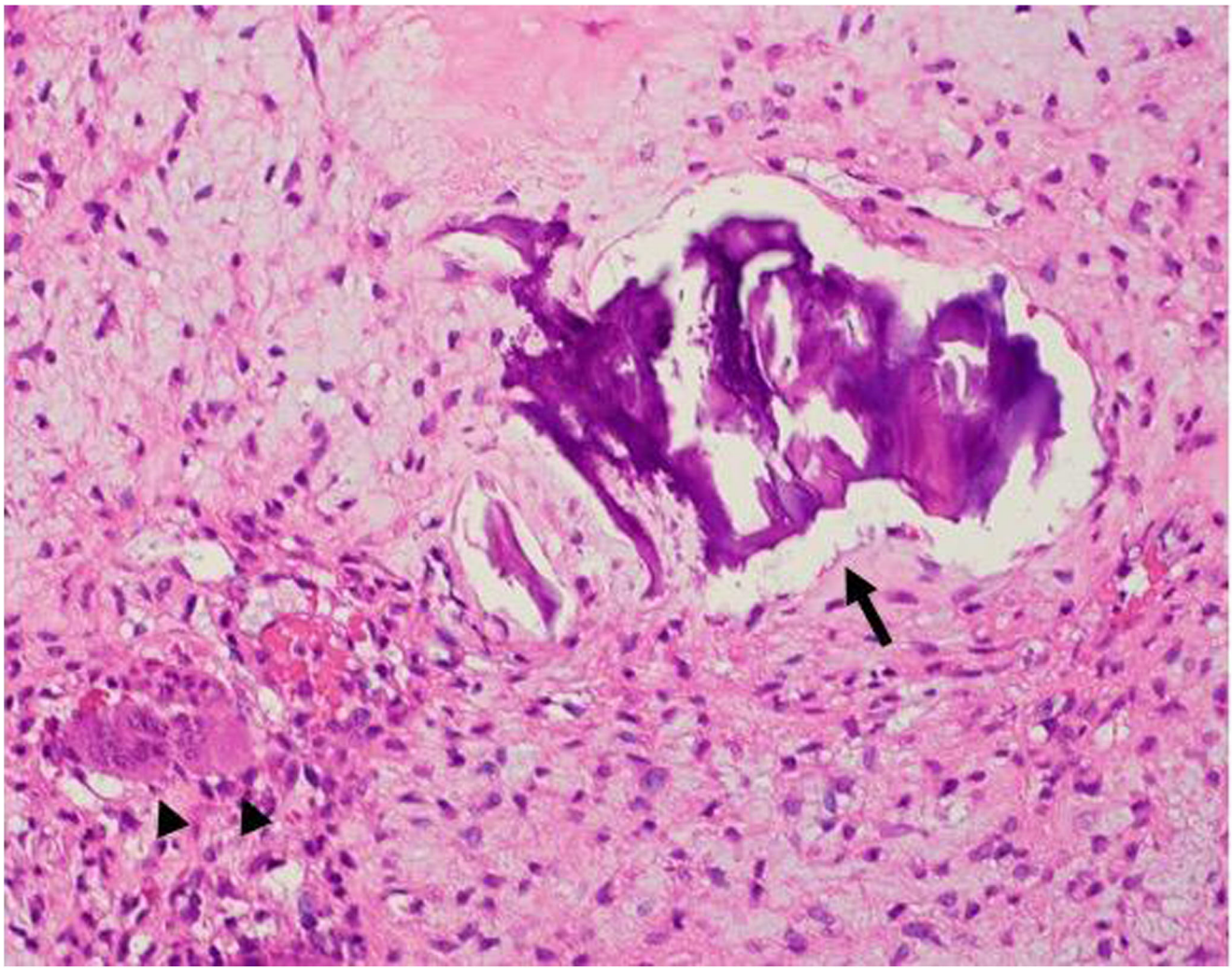

Fig. 6 High-power magnification demonstrates circumscribed calcification present near periphery (*arrow*) and adjacent to giant cell rich area (*arrowheads*)

described in the proximal tibia, women have a higher average age of 51 years, compared with 33 years in men. The demographics of the published surface-type CMF is summarized in Table 1.

Radiologically, the lesions tend to be small and very well circumscribed lucencies, with scalloping or saucerization of the underlying bone on radiography and CT. Some case reports describe maintenance of a cortical rim in lesions that were predominantly intracortical or subperiosteal [22]. The three cases described by Marin et al. all had disruption of the outer cortex [11]. Where described, all cases had an intact inner cortex and no intramedullary involvement. Interestingly, a proportion of cases reported punctate calcification, central calcification or a matrix, which is typically not a feature of conventional CMF in younger individuals [10, 11, 17]. Baker et al. noted that calcifications were not reported radiologically in their series; however, it was present microscopically within these lesions. In this case, faint chondroid-like calcification was appreciable on radiography.

MRI features of surface-type CMF were first reported by Marin et al. in 1997 and were included in all subsequent case reports. On MRI, the lesions are typically isointense to muscle on T1-weighted imaging and only one case noted the tumor to be mildly hyperintense [11]. On T2-weighted sequences, all lesions were hyperintense, with some demonstrating signal heterogeneity or central septa-like hypointensities [11, 12, 14, 16]. Enhancement patterns were infrequently reported, and those that were reported predominantly described either peripheral or heterogeneous enhancement [12, 13, 16, 17]. In this case, the enhancement pattern was diffuse and homogenous. Imaging findings are summarized in Table 1.

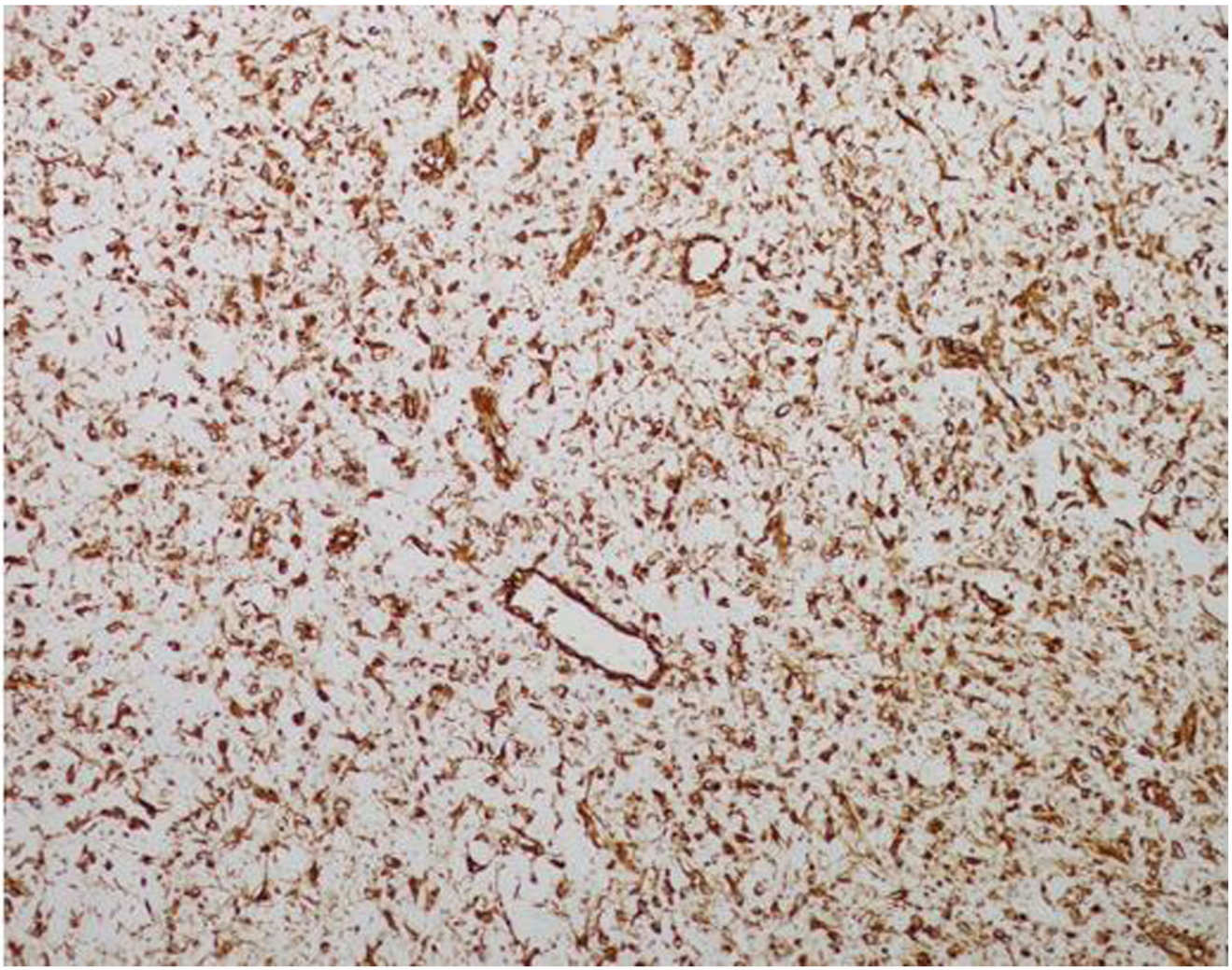

Fig. 7 Medium-power magnification demonstrates positive cell cytoplasmic staining for vimentin

Differential considerations of a well-circumscribed lucent-surface lesion of bone on imaging includes lesions, which may arise from the mesenchymal elements found there such as osteoid, cartilage, or fibrous tissue [23–25]. Bone-forming surface tumors such as osteoid osteomas may appear lucent with little surrounding sclerosis when formed in a joint space at the end of a bone [26]. Surface osteoblastomas may also appear lucent although they typically have marked surrounding soft tissue and bone edema. Juxtacortical chondroma is a cartilage forming tumor and can be viewed as a surface variant of the enchondroma. When they occur on the surface they are usually lucent and may contain chondroid calcification. They are typically found in the younger population with a predilection for the proximal humerus. Paraosteal chondrosarcoma is typically seen in the third or fourth decades of life and with a slight male predilection [27]. They tend to occur in long bones and can present as a well-circumscribed juxtacortical soft tissue mass with chondroid matrix. The underlying cortex may be normal, saucerized, or demonstrate cortical buttressing from a chronic periosteal reaction. Fibrous tumors, such as fibrous cortical defects, may be juxtacortical in location, although this tumor is typically a lesion of childhood. Non-neoplastic, post-traumatic lesions may also be included in the differential such as subperiosteal hematoma and periostitis ossificans, the latter may be associated with florid periosteal new bone [23]. Finally, parosteal ganglion is an entity typically described along the proximal tibia close to the pes anserinus insertion although other sites, including the femur, have been reported. They can cause erosion of the underlying cortical erosion as well as reactive bone formation. MRI is useful in confirming the cystic nature of the lesion [28].

Histologically, CMFs contain lobules of stellate and spindle-shaped cells associated with a myxoid, fibrotic, and chondroid matrix. These cells have nuclei that may vary from vesicular and ovoid to monochromatic, which can bear resemblance to

Table 1 Previously reported surface-type chondromyxoid fibromas and their imaging findings

Author	Year	Age	Sex	Bone (*n*)	Imaging features	MRI findings[a]
Jaffe	1958	"Young"	F	Humerus	Intracortical, lucent	Not performed
Ralph	1962	45	M	Fibula mid	Cortical lesion, lytic	Not performed
Andrew	1982	33	M	Proximal tibia	Periosteal reaction, soft tissue on cortical surface	Not performed
Bialik et al.	1985	5	F	Proximal femur	Internal calcification, periosteal reaction	Not performed
Schajowicz	1987	13	M	Proximal tibia	Intracortical, well circumscribed	Not performed
		30	M	Proximal tibia	Lucent, partially cortical	Not performed
		41	F	Proximal tibia	Intracortical, sclerotic rim	Not performed
Kenan et al.	1994	31	M	Proximal tibia	Cortical saucerization, periosteal reaction	Not performed
Park et al.	1995	16	F	Distal femur		Not performed
Marin et al.	1997	25	M	Proximal humerus	Punctate calcification, saucerization	T1 iso, T2 hyper
		45	M	Mid tibia	Lytic, cortical	T1 hyper, T2 hyper
		53	F	Proximal tibia	Expansile, sclerotic rim	T1 iso, T2 hyper
Park et al.	2000	20	M	Lesser trochanter	Well-defined sclerotic margin	T1 iso, T2 hyper, heterog enhance
Fujiwara	2003	25	F	Humerus	Well-defined, lucent, intact inner cortex	T1 iso, T2 hyper, heterog enhance
Baker et al.	2007	12–81	M&F	Proximal tibia [10]	Described in 2 patients only. Both were well circumscribed with cortical saucerization	Both lesions had only T1 features described, both were T1 iso.
		17–43	M&F	Distal femur [7]		
		64	M	Humerus		
		46	F	Rib		
		65	M	Metacarpal		
Takenaga	2007	36	M	Distal tibia	Lucent, saucerization, periosteal reaction	T1 iso, T2 hyper, enhances
		45	M	Distal fibula	Lytic, saucerization	T1 iso, T2 heterog hyper
Jhala et al.	2008	12	M	Proximal tibia	No radiograph	T1 iso, T2 hyper, periph enhance, perilesional marrow edema
Fernandez-Hernandez	2011	44	M	Proximal tibia	Lytic, sclerotic rim, matrix present	T1 iso, T2 hyper, heterog enhance
Abdelwahab	2013	41	F	Distal ulna	Lucent, well-defined sclerotic rim, no matrix,	T1 iso, T2 hyper with septae, periph enhance,
Soni et al.	2016	15	M	Distal femur	Lucent, sclerotic margins, septations	T1 iso, T2 hyper
Han et al.	2017	17	M	Metatarsal diaphysis	Intracortical, lytic, no matrix	T1 iso, T2 hyper with septae, periph enhance

[a] T1 iso = T1 isointense to muscle, T2 hyper = T2 hyperintense, heterog enhance = heterogeneous enhancement, homog enhance = homogenous enhancement, periph enhance = peripheral enhancement

those cells seen in chondrosarcoma. Scattered giant cells may also be observed intralesionally. Baker et al. note that histological features of surface-type CMF are almost identical to conventional CMF with some notable differences, the main one being the presence of foci of calcification within the chondroid matrix. This was seen to occur much more frequently than in the conventional CMF, where it only occurs in about one-third of cases. Reports of positive immunoreactivity for S-100 protein vary in the literature for both conventional CMF and surface-type CMF [2, 29]. In this case, S-100 was negative.

Conclusions

We reviewed 40 cases of surface-type chondromyxoid fibroma reported in the literature. There is a wide range of reported ages, from 6 to 87 years of age. Imaging findings are generally in keeping with a non-aggressive bone lesion, occasionally demonstrating chondroid matrix, and therefore juxtacortical chondroma frequently is the favored differential. A small number of published case reports have described a surface-type CMF with similar features to that of a juxtacortical

chondroma and while its rarity make it unlikely to feature in a list of differential considerations, awareness of the entity is important.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

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