Malignant transformation of a multiple cartilaginous exostosis – a case report

R. Willms, C.-H. Hartwig, P. Böhm, S. Sell

Orthopaedic Clinic, University of Tübingen, Tübingen, Germany

Accepted: 23 September 1996

Summary. *A patient is reported who had multiple cartilaginous exostoses with malignant transformation of a pelvic osteochondroma.*

Résumé. *On décrit le cas d'un patient atteint d'exostoses ostéocartilagineuses multiples avec dégénérescence maligne d'un ostéochondrome pelvien en chondrosarcome. On passe ensuite en revue 94 autres cas décrits par d'autres auteurs. L'incidence de la dégénérescence de la maladie ostéogénique exostotante est de 15%, l'âge moyen des patients étant de 31 ans. Les hommes sont deux fois plus touchés que les femmes. Plus de 85% des tumeurs étaient situées sur la partie proximale de l'humérus, sur le tronc ou sur la partie proximale du fémur. Le laps de temps entre le premier symptôme et l'établissement du diagnostic était en moyenne de deux ans et quatre mois. Une augmentation de taille était le symptôme majeur. Dans 9 cas sur 10, l'histologie révélait un chondrosarcome. Malgré les traitements à visées curatives, un patient sur deux faisait deux récurrences en moyenne. Chez 10% des patients des métastases étaient détectées. Un patient sur quatre avait une survie postopératoire de plus de 5 ans. Suit une discussion sur les déductions diagnostiques et thérapeutiques.*

Introduction

Malignant transformation of multiple cartilaginous exostoses is a serious complication [7, 19, 23]. A study of 59 patients reported malignant transformation in 46 [19]. There have been many reports giving basic details [1, 2, 6, 8, 9–12, 15, 17, 18, 23, 25], but in these malignancy was excluded when it was not proved histologically, although it was suspected on clinical grounds and radiographs. There are other cases where benign and malignant degeneration was seen consecutively [3, 5, 10, 20, 21, 23].

Our case is the only sarcoma arising from transformation of a cartilaginous exostosis which has been treated at our clinic in the last 10 years.

Case report

A man, 48 years of age, had a family history of osteochondroma, and had been known to have had tumours since the age of 12 years. For the previous 4 months, he had moderate abdominal pain, erectile dysfunction and urinary symptoms.

He had multiple exostosis affecting the proximal part of the right femur, both knee joints and the right distal tibia. There was a tumour the size of two clenched fists in the lower abdomen which could also be felt on rectal examination. Radiographs showed a lobulated, amorphous, calcified tumour.

A bone scan showed increased uptake in the pelvic area, and in both lower legs. A CT scan and MRI showed a tumour, 16×12×13 cm, arising from the left pubic bone which almost filled the lower part of the pelvis (Fig. 1). An intravenous pyelogram and a double contrast barium enema showed the bladder deformed into a dumb-bell shape which extended cranially and produced narrowing of the sigmoid colon.

Biopsy showed an undifferentiated chondrosarcoma. He had multiple metastases and was treated conservatively. He died from pulmonary metastases 6 weeks after the biopsy.

Fig. 1. **a** CT scan of the pelvis showing a lobulated amorphous, calcified tumour, 18 cm in diameter, arising from the left pubic ramus. **b** MRI

Discussion

The first report of malignant transformation of a cartilaginous exostosis was in 1886 [6], and since then we have been able to review 50 cases in the literature, 46 of which are included in Ochsner's survey [19]. These data are shown in Table 1 and Fig. 2. In 433 patients who had been operated on, malignant transformation was diagnosed in 67 (15%).

The risk of such transformation has been estimated at 3% on the basis of family analysis [11]. Garrison et al. operated on 24 malignancies in 88 patients (27%) [10].

Fig. 2. Diagram showing site of cartilaginous exostoses with malignant transformation in 96 cases from a review of the literature

Malignant transformation of osteochondroma occurred most frequently between the age of 19 and 39 years in contrast to the highest incidence of chondrosarcoma which is at 45 years [4, 16].

Two out of 3 malignancies in our review occurred in the trunk, and 88% occurred in these sites plus the proximal humerus and femur. In 76 patients with multiple cartilaginous exostoses, the commonest sites were the scapula, the proximal humerus, the pelvis and the proximal femur followed by the distal femur, the proximal or distal lower leg, the distal lower arm and the hand [22]. The low rate of malignant transformation of peripheral tumours may be because they are associated with compression syndromes or deformities and are removed at an earlier stage than tumours of the trunk.

Malignant transformation led to a chondrosarcoma in 90% of cases. Low rates of malignancy are reported [10]. Development to an undifferentiated chondrosarcoma has been described [6, 17] and this has a worse prognosis [4].

The large number of recurrences in spite of treatment and the short survival time may be because the patients seek advice late and operation was not sufficiently radical. The patients should be examined every 2 years while they are still grow-

Table 1. Summary of 96 cases from the literature of malignant transformation of multiple cartilaginous exostoses

Age (<i>n</i> = 95)	0–9 0	10–19 14	20–29 33	30–39 29	40–49 15	50–59 3	60–69 0	70–79 1
Sex (<i>n</i> = 95)	Male 66					Female 29		
Signs and symptoms (<i>n</i> = 43)	Tumour 43 (100%)			Pain 16 (37%)		Compression 8 (19%)		
Time since onset in years (<i>n</i> = 37)	< 1 12	1–5 20			6–10 4		> 10 1	
Histology (<i>n</i> = 96)	Chondro- sarcoma 90 (94%)			Osteo- sarcoma 4		Fibro sarcoma 1		Spindle-cell sarcoma 1
Therapy (<i>n</i> = 61)	Curative 50 (82%)			Palliative 11 (18%)				
Recurrences (<i>n</i> = 61)	Patients 25			Number of recurrences 47				
Metastases (<i>n</i> = 95)	Lung 6			Liver 1				
Survival-time in years (<i>n</i> = 39)	< 1 7 (18%)	≥1 24 (62%)			≥5 5 (13%)		≥10 3 (7%)	

ing, and after that every 3 to 5 years or whenever there are symptoms.

The radiological criteria of malignant transformation are an increase in size, increasing irregularity of the cartilage surface, development of a soft tissue tumour, and calcification outside the expected boundary of the tumour [14]. Exostoses with a diameter of more than 8 mm, and cartilaginous caps reaching more than 1 cm in adults, should be suspected of malignancy [24].

References

- Barnes R, Catto M (1966) Chondrosarcoma of bone. *J Bone Joint Surg [Br]* 48: 729–764
- Bennet GE, Berkheimer GA (1941) Malignant degeneration in a case of multiple benign exostoses. *Surgery* 10: 781–792
- Canella P, Gardini F, Boriam S (1981) Exostosis: development, evolution and relationship of malignant degeneration. *Ital J Orthop Trauma* 7: 293–298
- Dahlin DC (1978) Bone tumors. Thomas, Springfield
- Dorfmann HD (1973) Malignant transformation of benign bone lesions. *Proc Nat Cancer Conf* 7: 901–913
- Du YK, Shih HN, Wang JM, Shih CH (1991) Dedifferentiated chondrosarcoma arising from osteochondromatosis. *Chan Gung Med J* 14: 130–135
- Enderle A, Willert HG, Zichner L (1984) Angeborene und erworbene Skeletterkrankungen. In: Witt AN, Rettig H, Schlegel KF, Hackenbroch M, Hupfauer W (eds). *Orthopädie in Praxis und Klinik*. Thieme, Stuttgart New York
- Epstein DA, Levin EJ (1978) Bone scintigraphy in hereditary multiple exostoses. *Am J Roentgenol* 130: 331–333
- Fabaron F, Vandermarcq P, Pascard JP, Defaux F, Azaris O, Barret D, Drouineau J, Gasquet C (1990) Chondrosarcome intrathoracique à point de départ costal chez un patient porteur d'une maladie des exostoses multiples. *J Radiol* 71: 657–662
- Garrison RC, Unni KK, McLeod RA, Pritchard DJ, Dahlin DC (1982) Chondrosarcoma arising in osteochondroma. *Cancer* 49: 1890–1897
- Gordon ST, Buchanan JR, Ladda RL (1981) Hereditary multiple exostoses: report of kindred. *J Med Genet* 18: 428–430
- Hudson TU, Springfield DS, Spanier SS, Enneking WF, Hamlin DJ (1984) Benign exostoses and exostotic chondrosarcomas: evaluation of cartilage thickness by CT. *Radiol* 152: 595–599
- Huvos AG (1979) Bone tumors. Diagnosis, treatment and prognosis. Saunders, Philadelphia London Toronto
- Kotz R, Salzer-Kuntschik M, Lechner G, Immenkamp M (1984) Knochentumoren. In: Witt AN, Rettig H, Schlegel KF, Hackenbroch M, Hupfauer W (ed) *Orthopädie in Praxis und Klinik*. Thieme, Stuttgart New York
- Lange RH, Wisconsin M, Lange TA, Rao BK (1984) Correlative radiographic, scintigraphic and histological evaluation of exostoses. *J Bone Joint Surg [Am]* 66: 1454–1459
- Lichtenstein L (1972) Bone tumors. Mosby, St Louis
- Matsuno T, Ichioka Y, Yagi T, Ishii S (1988) Spindle-cell sarcoma in patients who have osteochondromatosis. *J Bone Joint Surg [Am]* 70: 137–141
- Middlemiss JH (1964) Cartilage tumours. *Br J Radiol* 37: 2277–2286

19. Ochsner PE (1978) Zum Problem der neoplastischen Entartung bei multiplen kartilaginären Exostosen. *Z Orthop* 116: 396–378
20. Schajowicz F (1981) Tumors and tumorlike lesions of bone and joints. Springer, New York Heidelberg Berlin
21. Shapiro F, Simon S, Glimcher MJ (1979) Hereditary multiple exostoses. *J Bone Joint Surg [Am]* 61: 815–824
22. Solomon L (1963) Hereditary multiple exostosis. *J Bone Joint Surg [Br]* 45: 292–304
23. Solomon L (1974) Hereditary multiple exostosis. *South African Med J* 48: 671–676
24. Spjut HL, Dorfman HD, Fechner RE, Ackerman LV (1971) Tumors of bone and cartilage. Armed Forces Institute of Pathology, Washington
25. Tsuchiya H, Morikawa S, Tomita K (1990) Osteosarcoma arising from a multiple exostosis lesion: case report. *Jpn J Clin Oncol* 20: 296–298
26. Weber O (1866) Zur Geschichte des Enchondroms namentlich im Bezug auf dessen hereditäres Vorkommen und sekundäre Verbreitung in inneren Organen durch Embolie. *Virchows Arch Pathol Anat* 35: 501–524