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Chondrosarcoma Arising in Polyostotic Fibrous Dysplasia

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As described by Coley and Stewart, malignant change of fibrous dysplasia is rare with incidence of 0.4 percent. A history of previous radiation therapy may or may not be associated the occurence. More malignancies have been reported as arising from polyostotic disease rather than from monostotic lesion. The most common tumor type is osteogenic sarcoma; less frequently fibrosarcoma, chondrosarcoma, and combination of these types have been described.

Authors report a case of chondrosarcoma arising from polyostotic fibrous dysplasia without history of previous radiation therapy. Review of the literature was made.

Case Report

The patient is a 32 year old male with chief complaints of pain and paresthesia on the left paranasal area and left nasal obstruction of one month duration. On admission, physical examination revealed exophthamos of the left eye, slight edema on left paranasal area, paresthesia of central area of the palate and marked hypertrophy of the inferior nasal turbinates. Routine hematology and urinalysis were within normal limits. Paranasal sinus series on roentgenography showed ill defined haziness on both maxillary sinuses and hypertrophy of chonca and septal deviation to the right side. Destruction of the left orbital floor was also seen and lateral view showed diffuse dense sclerotic change in skull base and facial bones (Fig. 1~3). Maxilloethmoidectomy was performed with tentative diagnosis of malignancy of the left maxillary sinus shortly after admission.

Gross findings: Round pinkish yellow hard mass measured approximately 3.8×2.8×2.0 cm in dimension. The cut surfaces showed multinodular trabeculated appearance intermingled with bony spicules.

Microscopic findings: The main component of the neoplasm was hyaline cartilage with single or multiple hyperchromatic and pleomorphic nuclei in a lacunar space. Mitotic figures were rare but nucleoli are prominent (Fig. 6, 7). The area of direct transformation of fibrous dysplasia to chondrosarcoma was observed (Fig. 8). Spindle cells were abundant enough to suspect fibrosarcoma in some areas (Fig. 9, 10). The specimen obtained by open biopsy at the left femur showed typical findings of fibrous dysplasia (Fig. 11).

Since the above microscopic findings indicated transformation of fibrous dysplasia to chondrosarcoma. Radiological study of bone series revealed typical changes of polyostotic

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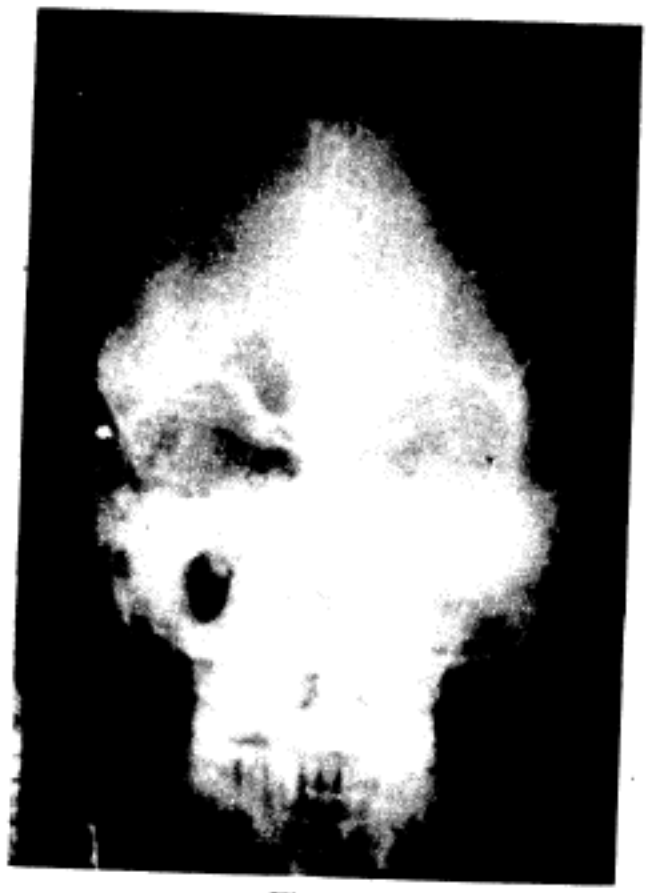

Fig. 1.

Fig. 2.

Fig. 3.

Fig. 1~3. (P.N.S. series)
There are noted ill-defined haziness in both maxillary sinuses, hypertrophy of chonca and septal deviation to the right side. In left maxillary lesion, suspicious destruction of the left orbital floor is also noted. In lateral view, diffuse dense sclerotic changes are noted at skull base & facial bones.

fibrous dysplasia at right first, 5 th, 7 th, 8 th and 10 th ribs, (Fig. 4, 5) pelvic bones, both femurs, humerus and tibias. Diffuse sclerotic changes of the skull base and facial bones also suggested leontiasis ossea. The symptoms of the patient have improved after the left maxilloethmoidectomy and he was discharged from the hospital. Four months after the operation, the maxillary mass has recurred and curretage was performed. Microscopically infiltrative, well differentiated chondrosarcoma were present. We noted areas

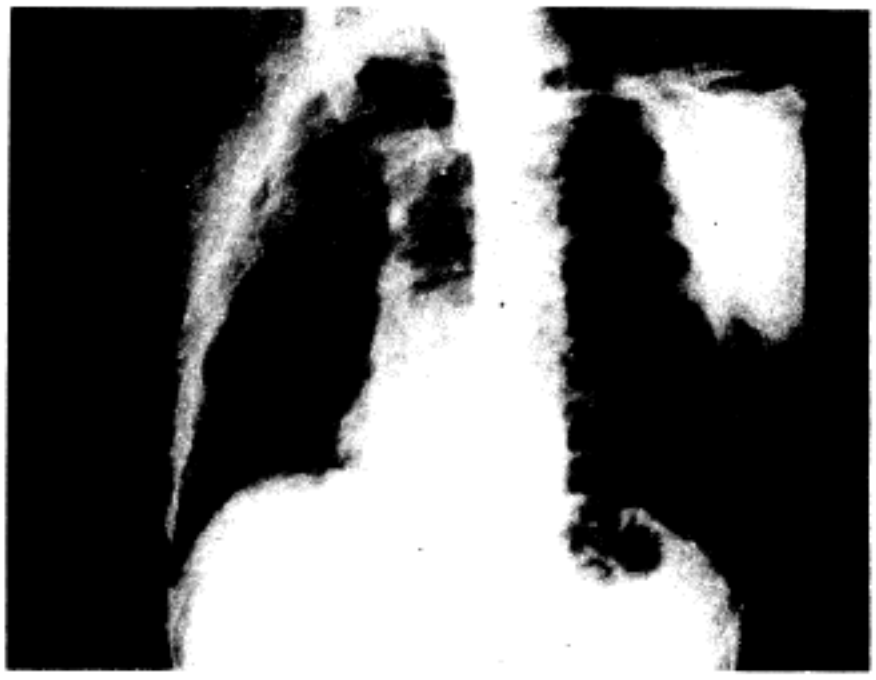

Fig. 4.

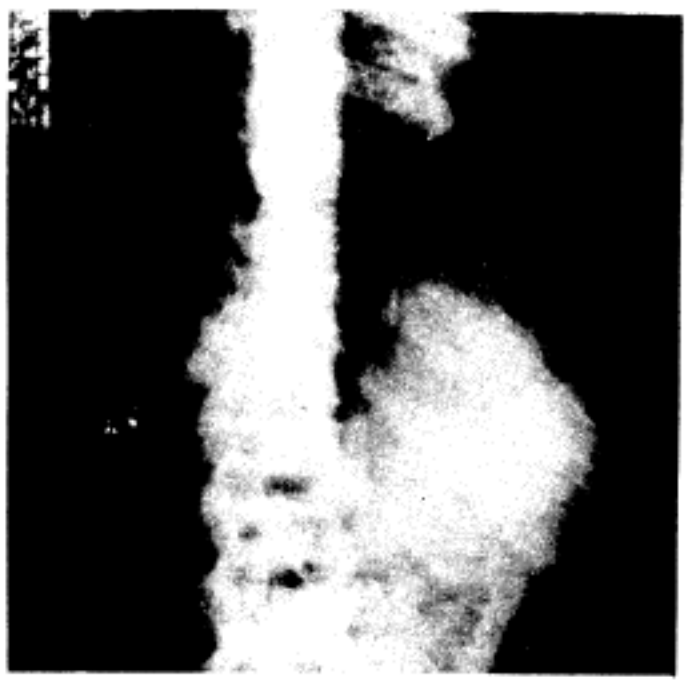

Fig. 5.

Fig. 4~5. (Rib cage study, LAO view)
There are noted soap bubble-like osteolytic expansile lesion in the right 5 th and 10 th ribs.

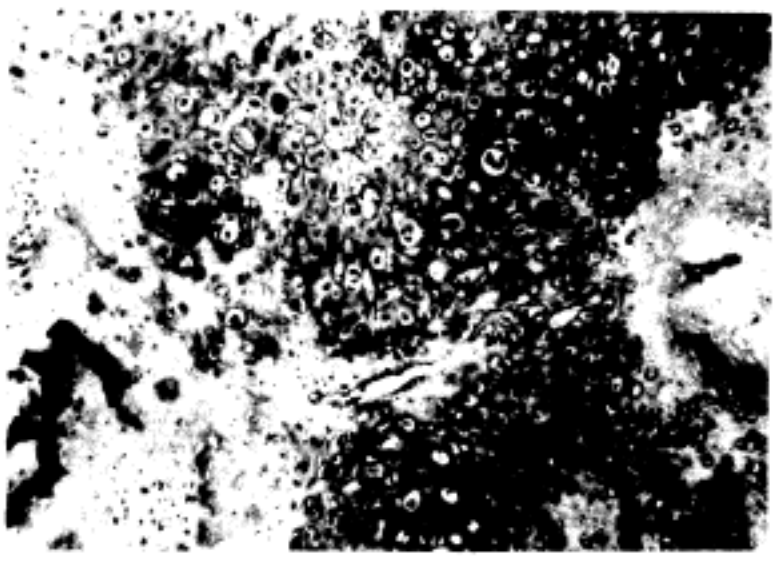

Fig. 6. Cartilage with single or multiple hyperchromatic & pleomorphic nuclei in single lacunar space.(H & E, ×100)

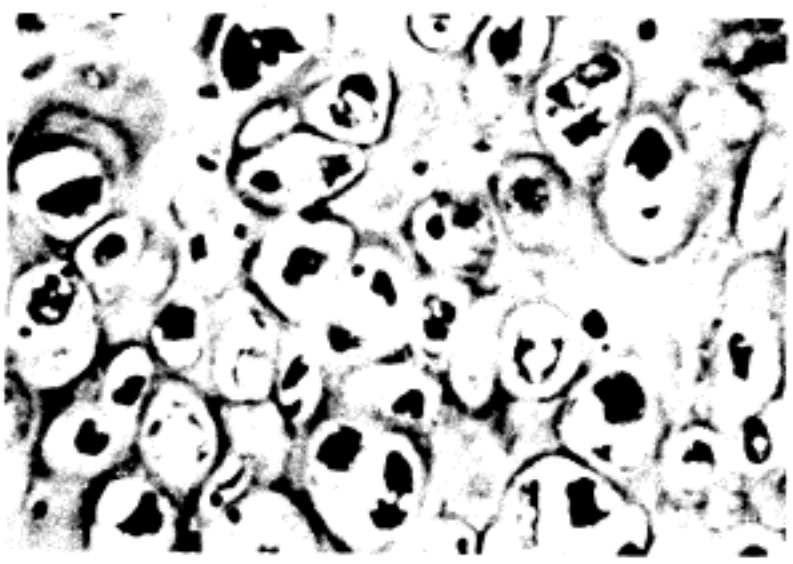

Fig. 7. High power view of Fig. 6. Anaplastic chondrocytes & multiple nuclei in single lacunar space(H & E, ×400)

of direct tranformation of fibrous dysplasia to chondrosarcoma (Fig. 8). Fibrous dysplasia was also conformed by an open biopsy of the bone at the trochanteric area of the left femur (Fig. 11). The patient was hopelessly discharged from the hospital.

Review and Dicussion

The etiology of fibrous dysplasia is entirely unknown. Lichtein[2~4] felt that the lesion is the result of perverted activity of bone-forming mesenchyme. Reed[5] thought arrest of bone maturation at woven bone stage. Knaggs and Philips suggested inflammation originated at dental alveoli, eyes, lacrimal sacs or paranasal sinuses as a fibrous dysplasia, and others suspected trauma as an etiologic factor[6,7].

The overall incidence of fibrous dysplasia is estimated to be 2.5 percent of all skeletal neoplasms and the incidence of malignant transformation is approximately 0.4 percent[3].

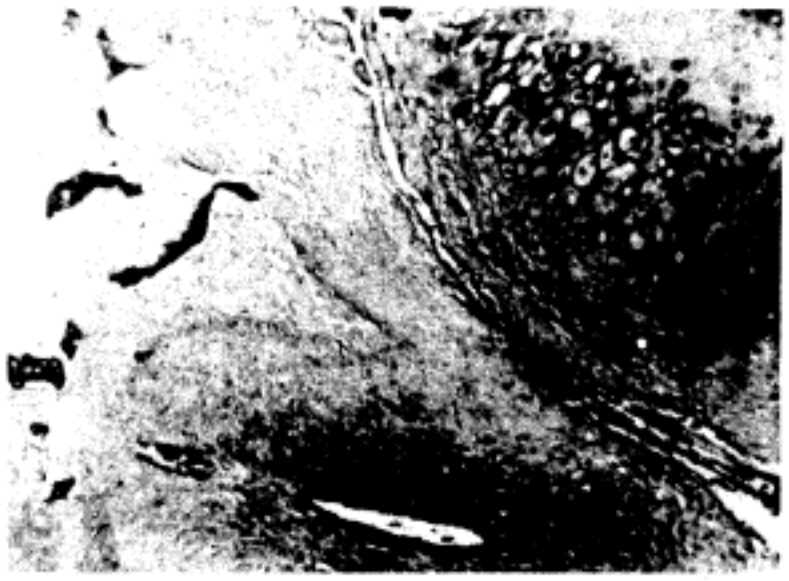

Fig. 8. The area of direct transformation of fibrous dysplasia(left) to chondrosarcoma. (H & E, ×40)

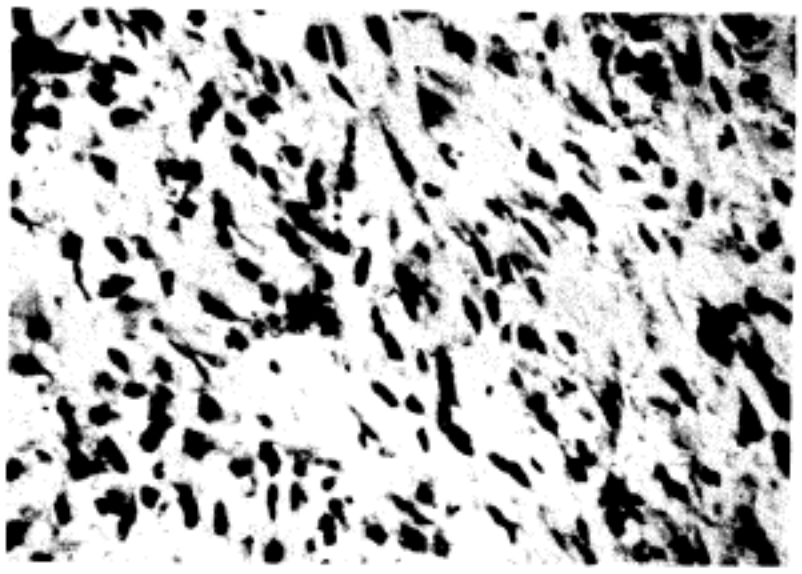

Fig. 10. High power view of Fig. 9 Spindle cells are abundant enough to suspect fibrosarcoma.(H & E, ×200)

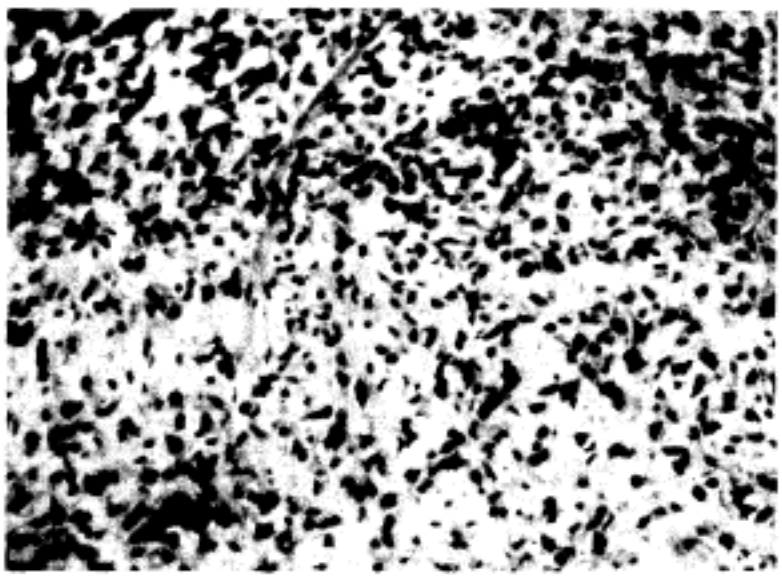

Fig. 9. Fibrosarcomatous portion.(H & E, ×100)

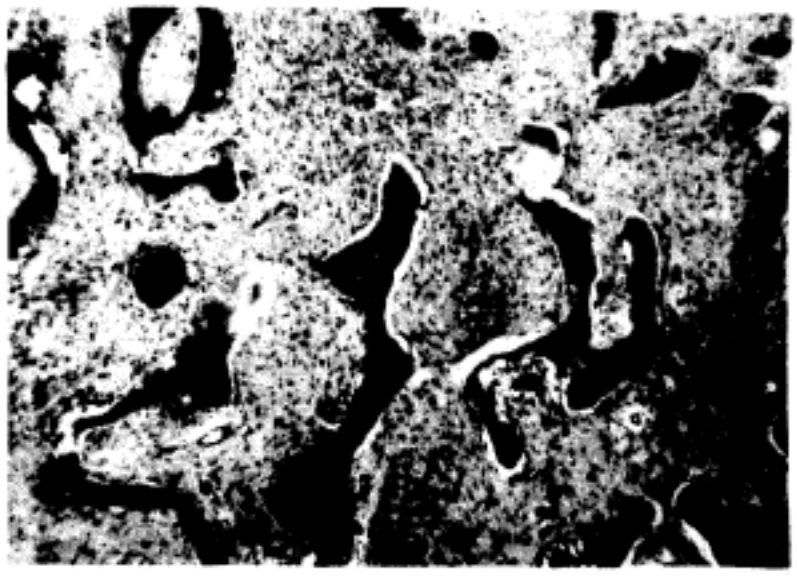

Fig. 11. Findings of typical fibrous dysplasia. (H & E, ×40)

Malignant changes are more common in polystotic fibrous dysplasia rather than in monostotic fibrous dysplasia. The most common tumor type is osteogenic sarcoma: less frequently fibrosarcoma, chondrosarcoma, or combination of these types have been described (Table 1, 2). Since the first description of malignant change of fibrous dysplasia by Coley and Stewart in 1945, Schwartz and Alpert collected 28 cases by 1964[1,3,9,10~15,17,18] From that time, numerous new cases have been reported[16,19,20~24,26~29], some of which developed without prior irradiation[30]. Among the 47 cases reviewed by the author from the literatures mentioned the above, polyostotic fibrous dysplasia was 26, and monostotic fibrous dysplasia was 21 cases. This indicates slightly increased incidence of occurrence in the polyostotic lesions. In these cases osteogenic sarcoma 27, fibrosarcoma 8, giant and spindle cell sarcoma 6, chondrosarcoma 5 and polymorphous osteoblastoclastoma was one case (Table 1, 2).

Onset of age for fibrous dysplasia varied from shortly after birth to 39 years with mean age of 16 years and onset of age for sarcoma in fibrous dysplasia was 8 to 61 years

Table 1. Previously repoted cases of malignant change in Fibrous dysplasia(1)

Year	Authors	Sex	Fibrous dysplasia			Sarcoma				
			Age at onset	Type of dysplasia	Affected bone	Age at onset	Affected bone	Histologic type	Meta-stasis	Result
1914	Elmslie			Polyostotic			Femur	Giant cell sar.		Dead
1927	Wanke[18]	F	7	Monostotic	Maxilla	37	Maxilla	Spindle cell sar	?	?
1928	Fromme	M	38	Monostoic	Humerus	43	Humerus	Osteogenic	+	Dead
1945	Coley & Stewart[1]	F	?	Polyostotic	Ribs, Pelvis, Femur, Skull	42	Scapula	Osteogenic	+	Dead
		M	?	Monostotic	Pelvis, Femur, Tibia	34	Femur	Giant cell sar.	+	Dead
1949	Belloni & Zanetti	F	16	Polyostotic	Femur. Hemipelvis	27	Femur	Osteogenic	+	Dead
	Shapper	F	7	Polyostotic	Femur, Humerus	14	Femur	Osteogenic	+	Dead
1950	Dustin & Ley[9]	F	7	Polyostotic	Femur, Tibia, Humerus, Skull, Hand, Pelvis	13	Pelvis	Osteogenic	+	Dead
1951	Sutro & Cabitza[39]	M	25	Polyostotic	Tibia. Fibula, Femur	30	Tibia	Osteogenic	?	Alive
		M	36	Polyostotic	Tibia, Metatarsal	39	Metatarsal	Chondrosarcoma	?	?
1955	Hall, Bersack & Vitolo[40]	M	32	Monostotic	Tibia	37	Tibia	Fibrosarcoma	?	?
	Pokinson & Higinbotham[41]	M	14	Polyostotic	Rib, Skull, Hemipelvis	22	Femur	Osteogenic	+	Dead
1956	Hobbs, Fischer & Back[11]	M	20	Monostotic	Skull	48	Frontal	Fibrosarcoma	0	Dead
	Portis	F	?	Polyostotic	Spine, Scapula, Humerus, Radius	39	Scapula	Osteogenic	+	Dead
	Sabanas et al[42]	F	6	Monostotic	Maxilla	24	Maxilla	Osteognic	+	Dead
	Trubnikov[43] &	M	39	Monostotic	Femur	61	Femur	Fibrosarcoma	0	Dead
	De Marchi	?	?	Monostotic	Mandible	?	Mandible	Osteogenic	0	Alive
1957	Parrini	F	6	Polyostotic	Maxilla, Mandible, Radius, Ulna, Hand, Calvarium	8	Calvarium	Osteogenic	?	?
1958	Jaffe[8]	F	7	Monostotic	Femur	20	Femur	Chondrosarcoma	0	Alive
	Vakhurkina[44]	M	30	Monostotic	Tibia	55	Tibia	Polymorphous osteoblastoclastoma	?	Dead

Table 1. Continued. (2)

Year	Authors	Sex	Fibrous dysplasia			Sarcoma				
			Age at onset	Type of dysplasia	Affected bone	Age at onset	Affected bone	Histologic type	Metastasis	Result
1961	Tanner, Dahlin & Childs	M	10	Monostotic	Maxilla	18	Maxilla	Osteogenic	?	Dead
		M	8	Polyostotic	Maxilla, Mandible	32	Mandible	Osteogenic	0	Alive
		M	22	Monostotic	Mandible	24	Mandible	Fibrosarcoma	+	Dead
1962	Haris Dudly & Barry[46]	M	4	Polyostotic	70% of skeleton	26	Femur	Osteogenic	+	Dead
	case 13 case 25	F	18	Polyostotic	20% of skeleton	49	Tibia	Myxofibrosarcoma	+	Dead
	Kieh, Deprez & Harris[47]	F	37	Polyostotic	Femur, Pelvis	42	Zygomatic	Osteogenic	0	Dead
	Sethi, Climie & Tuttle[48]	F	?	Monostotic	Rib	52	Rib	Osteogenic	+	Dead
	Jager[12]	F	12	Polyostotic	Maxilla, Clavicle, Ribs, Spine, Ulna, Radius, Femur, Tibia, Iliacs	22	Maxilla	Osteogenic	+	Dead
1963	Schwartz[2]	M	13	Monostotic	Mandible	20	Mandible	Fibrosarcoma	0	Dead
		F	10	Polyostotic	Craniofacial	24	Zygomatic	Osteogenic	+	Dead
	Van Horn, Hohnson & Dahlin[170]	?	?	Polyostotic	?	?	Femur	Osteogenic	+	Alive
1964	Riddell[27]	M	12	Polyostotic	Femur, Radius, Humerus	45	Femur	Spindle & Giant cell sercoma	0	Alive
1967	Gross & Montgomery[22]	F	19	Monostotic	Maxilla	50	Maxilla	Fibrosarcoma	+	?
1973	Feintugh[38]	F	Shortly after birth	Polyostotic	Femur, Humerus	40	?	Chondrosarcoma	0	?
1979	Johnson, Gilbert & Gottlieb[16]	F	2	Polyostotic	Femur, Tibia	25	Femur	Fibrosarcoma	+	Dead
1980*	Lim, Paik, Ko	M	32	Polyostotic	Crniofacial, Ribs, Femur, Humerus, Ulna	32	Maxilla	Chondrosarcoma	0	?

* Authors' case

Table 2. Bone sarcomas arising in fibrous dysplasia(Reported Cases by Huvos, Higinbotham & Miller[37], 1972

		Fibrous dysplasia			Sarcoma			
Case	Sex	Age at onset	Type of dysplasia	Bone aff cted	Age at onset	Bones affected	Histologic type	Result.
1	F	9	Polyostotic	Calvarium, maxilla, mandible.	34	Mandible	Osteogenic	Alive 3 years.
2	M	14	Polyostotic	Skull, ribs, ilium humisci	20	Femur	Osteogenic	Dead
3	F	9	Monostotic	Humerus	20	Humerus	Osteogenic	Alive 13 years
4	M	8	Monostotic	Femur	11	Femur	Chondrosarcoma	Alive 2 years
5	M	34	Monostotic	Mandible	49	Mandible	Osteogenic	Dead
6	M	24	Polyostotic	Pelvis, femur, tibia	34	Femur	Osteogenic	Dead
7	F	42	Polyostotic	Saull, rib, scapula, humerus, ilium, femur	42	Scapula	Spindle & giant cell sarcoma	Dead
8	F	19	Polyostotic	Ilium, ischium, pelvis	19	Pubis	Chondrosarcoma	Alive one year
9	M	17	Polyostotic	Femur, tibia	17	Femur, tibia	Osteogenic	Dead.
10	M	49	Monostotic	Fibula	49	Fibula	Osteogenic	Alive 4 years
11	M	54	Monostotic	Femur	54	Femur	Osteogenic	Alive 2 years
12	F	29	Monostotic	Mandible	29	Mandible	Spindle & giant cell sarcoma	Dead

* Cases 1 to 5 ; Existence of dysplasia was known years before the sarcoma arose
Cases 6 to 12 ; The discovery of the lesions was concurrent.

with mean age of 30.5 years. The lag period of fibrous dysplasia and sarcoma was two to fourty years with mean period of 14.8 years. The lag periods were 14.4 years for monostotic fibrous dysplasia and 15 years for polyostotic fibrous dysplasia. However, Schwartz and Alpert[3] reported lag period of monostotic fibrous dysplasia as 15 years and lag period of polyostotic fibrous dysplasia as 11.5 years, indicating earlier malignant change in polyos totic fibrous dysplasia.

Fibrous dysplasia associated with sarcoma in 47 reported cases, craniofacial bone and femur predominated with 16 cases each, tibia 5 cases, humerus, scapula, pelvis, 2 cases each, and fibula, rib, one case each. Schlumberger[31] reported chondrosarcoma arising more frequentlly in maxilla than in mandible, but authors have found 5 cases of maxilla and 7 cases of mandible in the review of the literatures.

Coley[32] argued that incidence of bone sarcoma is 0.001 percent in general population while incidence of malignant change of fibrous dysplasia is 0.4 percent. Therefore incidence of malignant change of fibrous dysplasia can be estimated to be 400 times higher than that of bone sarcoma in general population.

In 28 patient series reviewed by Stewart and Alpert, sarcomas always developed in bones affected by fibrous dysplasia and never in normal bones. Radiation may play a major role in pathogenesis of sarcoma. Many authors have claimed that the radiation given to the fibrous dysplasia is exclusively responsible for malignant change.

In Jaffe's experience of the sarcoma arising from an area of fibrous dysplasia, the fibrosarcoma had a better prognosis than that of the osteogenic sarcoma. However, in Schwartz's series, the 2 year survival rate was the same in both tumors (50%). The 5-year survival rate for the both was less than 20 percent. Whether the fibrous dysplasia was monostotic or polyostotic made no difference in the subsequent survival. The rate of metastasis was greater in cases developing after polyostotic fibrous dysplasia (90%) than monostotic fibrous dsplasia (50%). The most frequent site of metastasis is the lung.

Two other tumor types have been reported in patients with fibrous dysplasia. The adamantinoma (Baker Dockerty and Coventry[33], Cohen Dahlin and Pugh) and Cutaneous fibromyxoma (Braunwarth[34], Heinemann and Worth[35], Lick and Wiehweger[36] and Uehlinger) have been reported.

Areas of cartilage can rarely be seen in fibrous dysplasia although chondrosarcoma may arise from such a nodule in fibrous dysplasia[37]. A case of chondrosarcoma arising from a cartilaginous area of previously irradiated fibrous dysplasia was reported by Feintugh[38].

Summery

We have described a case of polyostotic fibrous dysplasia in a 32 year old male patient who had malignant transformation into a chondrosarcoma with feature of fibrosarcomatous pattern focally. In our review of the literatures sarcomas occurred more often in polyostotic fibrous dysplasia than in monostotic form. There was a higher frequency of malignant changes in male affected by fibrous dysplasia. The most important findings heralding the malignant transformation were pain, swelling of the affected areas and a noticable changes in the roentgenological appearance. The cranio-facial region is the most common site of these sarcomas. The lungs are the most common metastatic site and the osteo-

genic sarcoma is the predominant histologic type of the malignant change.

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=국문초록=

다골성 섬유성 이형성증(Polyostotic Fibrous Dysplasia)에서 발생된 연골육종 1예

인제의과대학 병리학교실

임인숙 · 백인기 · 고일향

섬유성 이형성증의 악성변화는 매우 드물며 골육종, 섬유육종, 연골육종과 다른 육종의 순으로 발생하는 것으로 기술되어 있다. 이 악성변화는 방사선 치료와 관련이 있는 경우도 있고, 방사선 치료와 관련이 없는 경우도 있다.

본예는 방사선 치료를 받지 않은 다골성 섬유성이형성증에서 이행된 좌측 상악동의 연골육종 1예를 관찰하였기에 보고한다. 환자는 32세 남자로 1개월간 좌측 비주위(perinasal area)에 동통과 감각이상(paresthesia), 좌측 비폐쇄를 주소로 내원하였다. 이학적 검사상 좌측 안구돌출, 좌측 비주위에 약간의 부종, 구개부위의 감각이상, 하비갑개의 현저한 비후가 있었다.

X-ray검사상 거의 전신의 골격에 다발성으로 섬유성 이형성증을 볼 수 있었고, 좌측 상악동에는 섬유성 이형성증의 육종성 변화를 볼 수 있었다. 이상의 소견으로 좌측 상악동 악성종양으로 생각하여 수술을 시행하였다. 병리조직검사 소견은 분화가 좋은 연골육종이었고 섬유성 이형성증에서부터 이행부위도 관찰되었다.

4개월후에 상악동 종양이 재발하여 currette 했으며 역시 분화가 양호하나 침윤성인 연골육종의 소견을 볼 수 있었고, 좌측 대퇴골에서 개방성 조직생검을 하여 섬유성 이형성증을 확인하였다.