FIBROUS DYSPLASIA OF THE SPINE WITH SARCOMATOUS TRANSFORMATION
A Case Report and Review of the Literature

Erol YALNIZ* **Turgay ER**** **Filiz ÖZYILMAZ*****

A fibrosarcoma is reported in the spine of a 53-year-old male with polyostotic fibrous dysplasia. There was no history of endocrine disturbances and no previous irradiation. Malignant transformation in fibrous dysplasia kis rare. A review of the literature reveals 101 cases of malignant degeneration occurring in fibrous dysplasia. We believe that this is the first report of sarcomatous change arising in an area of fibrous dysplasia in the spine.

Key words: *Fibrous dysplasia, fibrosarcoma, malignant transformation.*

Fibrous dysplasia is a common benign pathological condition characterized by fibro-osseous metaplasia. There are monostotic and polyostotic forms. Polpostotic fibrous dysplasia may be accompanied by skin pigmentation, endocrine disorders and precocious puberty, and entity known as Albright's (1) syndrome.

Although any bone can be affected, vertebral column involvement is uncommon, especially in the monostotic type (5, 13, 16, 18). Thoracic localization is even more unusual. Wright and Stoker (18) reported 9 cases of vertebral involvement in polpostotic fibrous dysplasia; only one of cases had a lesion in the thoracic region.

Malignant degeneration of fibrous dysplasia is a rare phenomenon. The most common malignant tumor is osteosarcoma (53.4 %) followed by fibrosarcoma (17.8 %) and chondrosarcoma (8.9 %).

CASE

A 53-year-old man complained of back pain and weakness. His blood count, ESR and alkaline phosphatase level were normal. No previous radiotherapy had been administered. Plain roetgenograms showed expansive and lytic lesions involving the thoracolumbar spine and multiple ribs. Computerized axial tomography and magnetic resonance imaging revealed a compressed spinal cord between T7 and T10. A biopsy of the left tenth rib exhibited typical histopatological changes of fibrous dysplasia. The patient underwent an exploration of his thoracic and lumber spine through a posterior approach. A decompressive laminectomy between T7 and T11 was done with transpedicular vertebral biopsy and posterior spinal instrumentation applied. Histological examination of the specimen from the vertebral body and posterior elements revealed fibrous dysplasia with features of fibrosarcoma. His symptoms were relieved six courses of chemotherapy (Adriamycin). Twenty-eight months after operation, he is still alive.

DISCUSSION

Malignant degeneration of fibrous dysplasia is a rare but well recognized complication. 101 cases have been reported (2, 3, 4, 6, 7, 8, 9, 10, 11, 12, 14, 17, 19). The incidence is estimated at 0.4% (6/1517) for fibrous dysplasia and 4% for Albright's syndrome (12).

There was no prevalence for either sex. Of the reported cases, 44 had monostotic fibrous dysplasia, 46 polpostotic form and 11 Albright's syndrome. The common sites were craniofacial bones (35.6%) femur (24.7%) and tibia (12.8%). Osteosarcoma was the most common type of sarcoma that developed in fibrous dysplasia (54 cases). The next common tumors were fibrosarcoma (18 cases) and chondrosarcoma (9 cases). Ebata et al. (4) reported a case of polpostotic fibrous dyslasia in which two types of malignant tumor arose, a chondrosarcoma and an osteosarcoma. Malignant degeneration usually developes in the third or fourth decade of life. The age of onset is 32 years and the lag between the development of fibrous dysplasia and sarcoma is an average 13.5 years (12).

Malignant neoplasms are uncommon complications of radiation therapy. Radiation-induced sarcomas of bone are estimated to develope in 0.035 % of patients treated by irradiation who survive 5 years (15). The role of radiation for malignant change in fibrous dysplasia has been discussed. In the review by Yabut et al. (19), 23 of 83 patients had prior radiation therapy. Chetty et al. 53) reported 3 cases of malignant neoplasms occurring in fibrous dysplasia involving facial bones. Two of them had been irradiated. the interval time between radiation therapy and the onset of the

* University of Trakya. Faculty of Medicine. Department of Orthopaedics and Traumatology, Edirne, Assistant Professor.
** Balta Limanı Bone Disease Hospital, ∂stanbul, Orthopaedic Surgeon.
*** University of Trakya. Faculty of Medicine. Department of Pathology, Edirne, Assistant Professor.

sarcoma was 11 years (12). In this study, 28 patients (27.7 %) had a history of irradiation as treatment for fibrous dysplasia. In twenty of them (71.4 %), tumor was an osteosarcoma. Although the sarcomas also occurred in nonirradiated patients, it seems that irradiation provokes the fibrous dysplasia to undergo sarcomatous change. For this reason radiotherapy should not be used for the treatment of fibrous dysplasia.

REFERENCES

1. Albright F., Butter A.M., Smith P.: Syndrome characterised by osteitis fibrosa disseminata, areas of pigmentation and endocrine dysfunction, with precocious puberty in females: Report of five cases. N. Eng. J. Med. 216: 727, 1937.
2. Blackwell J.B.: Mesenchymal chondrosarcoma arising in fibrous dysplasia of the femur. J. Clin. Pathol. 46: 961, 1993.
3. Chetty R., Kalan M.R., Kranold D.H.: Malignant transformation in fibrous dysplasia. A report of 3 cases. S.Afr. J. Surg. 28 (): 80, 1990.
4. Ebata K., Usami T., Tohnai I., Kaneda T.: Chondrosarcoma and osteosarcoma arising in polyostotic fibrous dysplasia. J. Oral Maxillofac. Surg. 50 (7): 761, 1992.
5. Ehara S., Kattapuram S.V., Rosenburg A.E.: Fibrous dysplasia of the spine. Spine. 17 (8):977, 1992.
6. Ishida T., Machinami R., Kojima T., Kikuchi F.: Malignant fibrous histiocytoma and osteosarcoma in association with fibrous dysplasia of bone. Report of three cases. Pathol. Res. Pract. 188 (6): 757, 1992.
7. Jose C.C., Benjamin C.S.: Osteogenic sarcoma arising in polpostotic fibrous dysplasia. A case report. australas. Radiol. 30(2): 134, 1986.
8. Mock D., Rosen I.B.: Osteosarcoma in irradiated fibrous dysplasia. J. Oral. Pathol. 15 (1): 1, 1986.
9. Mortensen A., Bojsen-Moller M., Rasmunsen P.: Fibrous dysplasia of the skull with acromegaly and sarcomatous transformation. Two cases with a review of the literature. J. Neurooncol. 7(1): 25, 1989.
10. Present D., Bertoni F., Enneking W.F.: Osteosarcoma of the mandibula arising in fibrous dysplasia. A case report. Clin. Orthop. 204: 238, 1986.
11. Roth A., Touret P., Rigault P.: Association of Ewing's sarcoma with fibrous dysplasia of the tibia. Report of one case. Rev. Chir. Orthop. Reparatrice Appar Mot. 71 (2): 133, 1985.
12. Schwartz D.T., Alpert M.: The malignant transformation of fibrous dysplasia. Am. J. med. Sci. 247: 1, 1964.
13. Serena S., Healey J.H., Huvos A.G.: Fibrous dysplasia of the second cervical vertebra. J. Bone Joint Surg. 72A: 781, 1990.
14. Taconis W.K.: Osteosarcoma in fibrous dysplasia. Skeletal Radiol. 17(3): 163, 1988.
15. Tountas A.A., Formasier V.L., Harword A.R., Leung P.M.K.: Postirradiation sarcoma of bone. A perspective. Cancer. 43: 182, 1979.
16. Troop J.K., Herring J.A.: Monostotic fibrous dysplasia of the lumbar spine. Case report and review of the literature. J. Pediatr. Orthop. 8(5): 599, 1988.
17. Witkin G.B., Guilford W.B., Siegal G.P.: Osteogenic sarcoma and soft tissue myxoma in a patient with fibrous dysplasia and hemoglobins J. Baltimore and S. Clin. orthop. 204: 245, 1986.
18. Wright J.F.S., Stoker D.J.: Fibrous dysplasia of the spine. Clin. Radiol. 39(5): 523, 1988.
19. Yabut S.M., Kenan S., Sissons H.A., Lewis M.M.: Malignant transformation of fibrous dysplasia. A case report and review of the literature. Clin. orthop. 228: 281, 1988.