Recurrent Chondromyxoid Fibroma of the Thoracic Spine 30 Years After Primary Excision

Case Report and Review of the Literature

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We report a case of late recurrence of chondromyxoid fibroma (CMF) arising in a thoracic vertebra in an 11-year-old male. This was treated by curettage, and 30 years later, the patient noticed shoulder pain and leg weakness. A recurrent mass appeared at the same site in the spinous process of T6. The histologic features of the recurrent tumor were similar to those of the primary lesion. A total of 38 cases of CMF of the vertebra have been reported. Only 3 of 38 previously reported vertebral CMF recurred. Tumors recurred 2 years after operation in 2 cases, and 7 years after operation in 1 case. *Int J Surg Pathol 9(4):323–329, 2001

Key words: chondromyxoid fibroma, thoracic spine, recurrence.

Chondromyxoid fibroma (CMF) was first described by Jaffe and Lichtenstein [1] in 1948. They described 8 cases with tumor arising in the tibia (3 cases), femur (2 cases), metatarsal bone (2 cases), and calcaneus (1 case). CMF is a rare skeletal neoplasm. It accounts for less than 1% of all benign and malignant tumors of bone. The tumor has a predilection for males, with a male/female sex ratio of about 1.5:1, and commonly occurs in the second and third decades of life. The metaphyseal regions of major tubular bones such as the tibia and femur, small bones of the feet, and pelvic bone are the most frequent sites of involvement [2]. CMF of the spine is rare. Zillmer and Dorfman [3] reported that vertebral involvement occurs in about 8% of CMF cases. Wu et al. [4] reported 10% in 1998. Overall recurrence rates of CMF were reported by Dorfman and Czerniak [2], Wu et al. [4], and Gherlinzoni et al. [5] are 20%, 26.3%, and 27%, respectively.

Case Report

The patient was a 41-year-old man who presented at age of 11 years with a radiolucent lesion in the spinous process of the 6th thoracic vertebra. The tumor was excised from a “cystic” lesion of the spinous process. Dr. Henry L. Jaffe at the Hospital for Joint Disease in New York City was consulted, and the pathologic diagnosis was confirmed as chondromyxoid fibroma. The patient remained asymptomatic until 30 years later, when he presented with shoulder pain and leg weakness. A computed axial tomography (CAT) scan revealed an expansile and irregularly shaped lytic lesion with sclerotic borders involving the posterior elements of the 6th thoracic vertebra, predominantly the left pedicle and spinous process. The spinal cord at this level was displaced to the right and slightly compressed (Fig. 1). At the level of T6, axial magnetic resonance images
Fig. 1. Axial CT scan shows recurrent tumor of the 6th thoracic vertebra involving the left side of the posterior neural arch.

(MRI) showed a low-signal-intensity lesion with high-signal-intensity foci in the posterior vertebra (Fig. 2). Sagittal MRI showed an ill-defined and high-signal-intensity lesion in the posterior vertebral arch (Fig. 3). The patient underwent surgical excision of the recurrent lesion.

The resected tissue consisted of several fragments measuring up to $2.4 \times 1.8 \times 0.4$ cm. After formalin fixation, routine histopathological sections were made and stained with hematoxylin and eosin. Slides of the original specimen removed from the 6th spinous process of the thoracic vertebra were still available for review, but none of the original radiographs had been retained.

**Histologic Findings**

The lesion was predominantly composed of immature myxoid tissue with a pseudolobular appearance. Myxoid areas consisting of stellate and plump spindle cells alternating with areas of solid cellularity with plump, oval mononuclear cells and multinucleated giant cells were present. The tumor cells showed moderate nuclear hyperchromatism. Occasionally, large, bizarre hyperchromatic nuclei were present. No mitotic figures were found. Scant reactive bone formation was seen at the periphery (Fig. 4). The histologic features of the recurrent tumor were similar to those of the primary tumor lesion excised 30 years earlier, except for the presence of a focus of necrosis in the primary lesion (Fig. 5).

**Discussion**

The first report of occurrence of CMF in the vertebral column was presented by Benson and Bass [6] in 1955. They reported CMF of thoracic vertebral bodies (T1, 2, 3) of a 34-year-old woman.
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Fig. 4. Histologic features of primary tumor of the 6th thoracic vertebra (1969). A. Low-power photomicrograph shows loose fibromyxoid tissue and peripheral zone of high cellularity with several scattered multinucleated giant cells. B. High-power photomicrograph shows stellate and spindle cells in myxoid stroma.

Fig. 5. Recurrent tumor tissue of the 6th thoracic vertebra (1999). A. Low-power photomicrograph shows pseudolobulation with greater cellularity at periphery of fibromyxoid lesion. B. Higher magnification of the peripheral area of pseudolobule shows stellate and spindle cells, and several multinucleated giant cells.

Curettage and postoperative radiation therapy were performed. Fifteen months after the operation, the patient had no recurrence. Only 38 cases of vertebral CMF have been reported so far. These cases involved cervical vertebrae in 7 cases [3,4,7–10], thoracic vertebrae in 14 cases [3,4,6,11–17], lumbar vertebrae in 7 cases [4,18–21], and sacrum in 7 cases [3,4,22–25]. Precise locations were not described in 3 cases [26,27] (Table 1).

The ages and sexes of the patients were available for 23 of 39 cases, including our case. The patients in 23 cases ranged in age from 6 to 58 years (average age, 26.0 years); 78% of cases occurred in the second, third, and fourth decades. The 23 tumors occurred in 9 male and 14 female patients. The sex ratio was approximately 0.6:1. Zillmer and Dorfman and Wu et al. reported that all CMF patients, including spinal CMF, ranged in age from 3 to 70 years [3] and 6 to 87 years [4], respectively (average age, 31.1 years). Peak incidence was in the second and third decades of life [3,4]. Dorfman and Czerniak [2] stated that the male/female sex ratio was approximately 1.5:1.

The average age of patients with spinal CMF was younger than that of patients with CMF in general. The sex ratio was different in cases of spinal CMF, when compared with all cases of CMF. There was a female predominance in spinal CMF cases.
Follow-up data, including our case, were mentioned in 17 of 39 cases (Table 1). The range of follow-up data was from 2 months to 8 years after operation. Thirteen of 17 cases had no evidence of recurrence after operation; 4 of 17 cases, including our case, had recurrence. The first recurrent case (case no. 11) was reported by Nunez et al. [14]. The patient was a 38-year-old woman who had a tumor of the 5th thoracic vertebra with a compression fracture. Two years after curettage, she had recurrence and a transthoracic excision was performed. She had no evidence of recurrence 9 months after reexcision. The second recurrent case of spinal CMF (case no. 16) was reported by Zillmer and Dorfman [3]. The patient was a 20-year-old woman who had a tumor of the 7th cervical vertebra. Intralasional excision and radiation therapy were performed. Seven years later she had a tumor containing recurrent CMF and malignant fibrous histiocytoma. The latter was considered to be a radiation-induced tumor. A third recurrent case of CMF of spine (case no. 21) was reported by Leal Filho et al. [16]. The patient was a 32-year-old woman who had a tumor of the 5th costovertebral junction with spread into the spinal canal. Laminectomy and removal of an epidural tumor were performed. Two years after operation, she had recurrent tumor, which was resected. Two years after the second operation, there was no further evidence of recurrent tumor. It is interesting that 3 of 4 recurrent cases of spinal CMF, including our case, arose in thoracic vertebrae.

Review of the literature shows the lag period before the first recurrence of CMF in all sites ranged between 0.5 year and 9 years (Table 2) [28-44]. The longest interval before recurrence was 18 years in second recurrence of metatarsal CMF [41]. Zillmer and Dorfman [3] mentioned that the overall time to recurrence was less than 2 years, and Wu et al. [4] reported that it was between 5 months and 10 years (average age, 3.2 years), and the first recurrence was usually noted within 1 year. The 30-year interval before recurrence in our case is the longest yet reported.

The sex ratio of recurrent cases was 1:1.5, with 10 males and 15 females (Cases no. 11, 16, 21, 27 in Tables 1 and 2). The average age of these cases was 21.9 years, and 44% of the patients were younger than 15 years of age.

Ralph [45] stated that CMF was more aggressive in younger patients. He noted a higher local recur-

| Case No./Author(s) | Age/Sex | Location | Follow-up |
|--------------------|---------|----------|-----------|
| 1. Benson and Bass [6], 1955 | 34/F | T1,2,3 | 15 months, NED |
| 2. Gudsha [18], 1968 | 23/F | L3 | 6 months, NED |
| 3. Schajowicz [7], 1971 | 6/F | C3 | — |
| 4. Spjut et al. [26], 1971 | — | Vertebral (2 cases) | — |
| 5. Rahimi et al. [27], 1972 | 34/M | T12 | — |
| 6. Raja-Reddy et al. [11], 1973 | 44/M | T10 | — |
| 7. RamanI [12], 1974 | 9/M | L4 | 2 months, NED |
| 8. Tsuji et al. [19], 1975 | 11/M | T10 | 4 years, NED |
| 9. Merli et al. [13], 1978 | 23/M | L2 | 20 months, NED |
| 10. Mayer [20], 1978 | 38/F | T5 | Recurrence 2 years after operation |
| 11. Nunez et al. [14], 1982 | 20/F | C7 | 15 months, NED |
| 12. Standefer et al. [8], 1982 | — | S | — |
| 13. Makley et al. [22], 1982 | 15/M | S | 2 years, NED |
| 14. Shulman et al. [23], 1985 | 32/M | C4 | 2 years, NED |
| 15. Provelegios et al. [9], 1988 | 20/F | C7 | 7 years after operation, recurrence with MFH |
| 16. Zillmer and Dorfman [3], 1989 | 36/F | T10 | — |
| 17. Zillmer and Dorfman [3], 1989 | 58/F | S1,2 | — |
| 18. Zillmer and Dorfman [3], 1989 | 41/F | C5 | 10 months, NED |
| 19. Rivierez et al. [10], 1991 | 19/F | T2 | 2 years, NED |
| 20. Tsuichia et al. [15], 1992 | 32/F | T5 | Recurrence 2 years after operation |
| 21. Leal Filho et al. [16], 1995 | 19/F | L1,2 | 5 years, NED |
| 22. Cabral et al. [21], 1997 | 17/F | S | 8 years, NED |
| 23. Rodgers et al. [24], 1997 | — | C (2 cases) | — |
| 24. Wu et al. [4], 1998 | — | T (5 cases) | — |
| 25. Bruder et al. [17], 1999 | 27/F | T5 | 1 year, NED |
| 26. Brat et al. [25], 1999 | 30/M | S2,3,4 | 1 year, NED |
| 27. Present case, 2000 | 11/M | T6 | Recurrence 30 years after operation |

NED = no evidence of disease.
Table 2. Cases of Recurrent CMF

| Case No./Author(s) | Age/Sex | Location       | Follow-up                                      |
|-------------------|---------|----------------|-----------------------------------------------|
| 1. Jaffe [28], 1958 | 13/M    | Calcaneus      | One recurrence from 1st to 2nd operation, 9 months |
| 2. Iwata, Coley [29], 1958 | 17/M    | Fibula         | Two recurrences from 1st to 2nd operation, 1 year, from 2nd to 3rd operation, 6 months |
| 3. Crabbe [30], 1962 | 11/M    | Tibia          | Two recurrences from 1st to 2nd operation, 6 months, from 2nd to 3rd operation, 1 year |
| 4. Mikulowski, Ostberg [31], 1971 | 42/M    | Tibia          | Two recurrences from 1st to 2nd operation, 9 years, from 2nd to 3rd operation, 10 years |
| 5. Browne, Rivas [32], 1977 | 13/M    | Mandible       | One recurrence from 1st to 2nd operation, 2 years |
| 6. Kyriakos [33], 1979 | 33/F    | Femur          | Two recurrences from 1st to 2nd operation, 1 year 10 months, from 2nd to 3rd operation, 3 years 6 months |
| 7. Heydemann et al. [34], 1985 | 12/F    | Tibia          | Two recurrences from 1st to 2nd operation, 2 years, from 2nd to 3rd operation, 2 years |
| 8. Kreicbergs et al. [35], 1985 | 11/M    | Humerus        | One recurrence from 1st to recurrence 6 months, follow-up without 2nd operation |
| 9. van Horn, Lemmens [36], 1986 | 11/F    | Calcaneus      | Three recurrences from 1st to 2nd operation, 2 years, from 2nd to 3rd operation, 1 year, from 3rd to 4th operation, 1 year, from 2nd to 3rd operation, 7 years |
| 10. Danielsen et al. [37], 1991 | 22/F    | Mandible       | One recurrence from 1st to 2nd operation, 3 years |
| 11. Campus Filho et al. [38], 1992 | 23/F    | Tibia          | Two recurrences from 1st to 3rd operation unknown, from 3rd to 4th operation, 3 years |
| 12. Campus Filho et al. [38], 1992 | 33/F    | Periacetabular | One recurrence from 1st to 2nd operation, 1 year, 2 months |
| 13. Campus Filho et al. [38], 1992 | 19/M    | Tibia          | One recurrence from 1st to 2nd operation, 2 years |
| 14. Lingen et al. [39], 1993 | 10/M    | Mandible       | One recurrence from 1st to 2nd operation, 3 years |
| 15. Troncoso et al. [40], 1993 | 14/F    | Tibia          | Two recurrences from 1st to 2nd operation, 4 years, from 2nd to 3rd operation, 13 years |
| 16. O’Connor et al. [41], 1996 | 14/F    | Metatarsal      | Four recurrences from 1st to 2nd operation, 2 years, from 2nd to 3rd operation, 9 months, from 3rd to 4th operation, 3 years, from 4th to 5th operation, 4 years |
| 17. O’Connor et al. [41], 1996 | 9/F     | Phalanx        | One recurrence from 1st to 2nd operation, 1 year, 6 months |
| 18. O’Connor et al. [41], 1996 | 18/M    | Metatarsal      | Two recurrences from 1st to 2nd operation, 1 year, from 2nd to 3rd operation, 18 years |
| 19. Keel et al. [42], 1997 | 66/F    | Sphenoidoccipital bone | One recurrence from 1st to 2nd operation, 6 months |
| 20. Patino-Cordoba et al. [43], 1998 | 41/F    | Clivus         | One recurrence from 1st to 2nd operation, 1 year |
| 21. Shek et al. [44], 1999 | 16/F    | Skull base     | Three recurrences from 1st to 2nd operation, 4 years, from 2nd to 3rd operation, 3 years, from 3rd to 4th operation, 3 years |

Rerence rate after initial curettage in young children, particularly in those under the age of 15 years. On the other hand, Gherlinzoni et al. [5] stated that the recurrence rate had nothing to do with the age of the patient. They reported equal rates of recurrence above and below the age of 20 years [5]. In 1998, Wu et al. [4] reported that the average age of patients with recurrent CMF was 22.6 years, and 40.6% of the patients were younger than 15 years of age.

Zillmer and Dorfman [3], Gherlinzoni et al. [5], and Schajowicz and Gallardo [7] stated that the histologic findings had no prognostic significance. In our case, there is no conspicuous difference in the histologic features in the primary and the recurrent lesions.

Conclusion
We report a case of CMF arising in a thoracic vertebra with recurrence, 30 years after the initial operation. CMF arising in the vertebral column is rare. Extensive review of the literature showed this case was the only instance in which the lag period before recurrence was as prolonged as 30 years. This case indicates the need for long-term follow-up in cases of CMF.
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