

Case Report

Monostotic fibrous dysplasia involved in cervical vertebrae

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Abstract: Fibrous dysplasia (FD) was firstly reported by Lichtenstein in 1938 and can be divided into two main kinds of subtypes: monostotic fibrous dysplasia (MFD) and polyostotic fibrous dysplasia (PFD) with or without endocrinopathy. MFD involved in cervical vertebrae is extremely rare. Two special cases of MFD involved in cervical vertebrae were treated by anterior surgical resection and fusion with instrumentation. In addition, the relevant literatures were reviewed and the characteristics of 13 MFD cases (including two case in our study) involved in cervical vertebrae were also summarized. Neither clinical symptoms nor signs of tumor recurrence were detected in our two cases at the last follow-up. The main presenting symptoms were neck pain, posttraumatic, pathologic fracture, painful torticollis, and incidental finding. The treatment methods included: biopsy/observation, corpectomy with instrumented fusion, posterior fusion, vertebroplasty, curettage and bone graft and complete removal with a combined anterior and posterior fusion procedure. Outcomes of these patients have been uniformly good with a mean follow-up time of 3.65 years (six months to 20 years). Histopathological examination is critical for the diagnosis of FD. Whether, when and how to treat a FD involved in cervical vertebrae still remains controversial. Surgical intervention is indicated if there is a mechanical reason for the pain or a cervical instability or even a compromise of neurological structures. There is no gold standard in operative treatment method and each treatment should be individualized.

Keywords: Monostotic fibrous dysplasia, surgical resection, cervical spine, literature review

Introduction

Fibrous dysplasia (FD), firstly reported by Lichtenstein in 1938, is a bone formation disorder characterized by the replacement of bone and marrow with poorly organized spicules of immature bone in a fibrous connective tissue [1]. FD can be divided into two kinds of subtypes: monostotic fibrous dysplasia (MFD) and polyostotic fibrous dysplasia (PFD) with or without endocrinopathy. During the past two decades, less than 40 cases of spinal FD with a limited follow-up duration have been reported according to a review published in 2013 [2]. The natural history of spinal FD has not been fully understood, and its treatment strategies (biopsy and/or observation, surgical resection, vertebroplasty and balloon kyphoplasty) still remain controversial. Considering the little knowledge of this area, we present two special cases of MFD involved in cervical vertebrae treated by anterior surgical resection and fusion with instrumentation to share our experience. In

addition the relevant literature were reviewed and the characteristics of previous MFD cases involved in cervical vertebrae were also summarized.

Case report

The patients provided informed consents for the publication of his or her clinical and radiological data. This study was approved by Medical Ethical Committee of West China Hospital, Sichuan University.

Case 1

A 21-year-old male patient presented to our hospital in June 2013 with chief complaint of persistent neck pain for 12 months. He had no symptoms of numbness, weakness, and abnormal sensation. Neurological examination on admission revealed no obvious abnormality. Three-dimensional computed tomography (CT) scan and magnetic resonance imaging (MRI)

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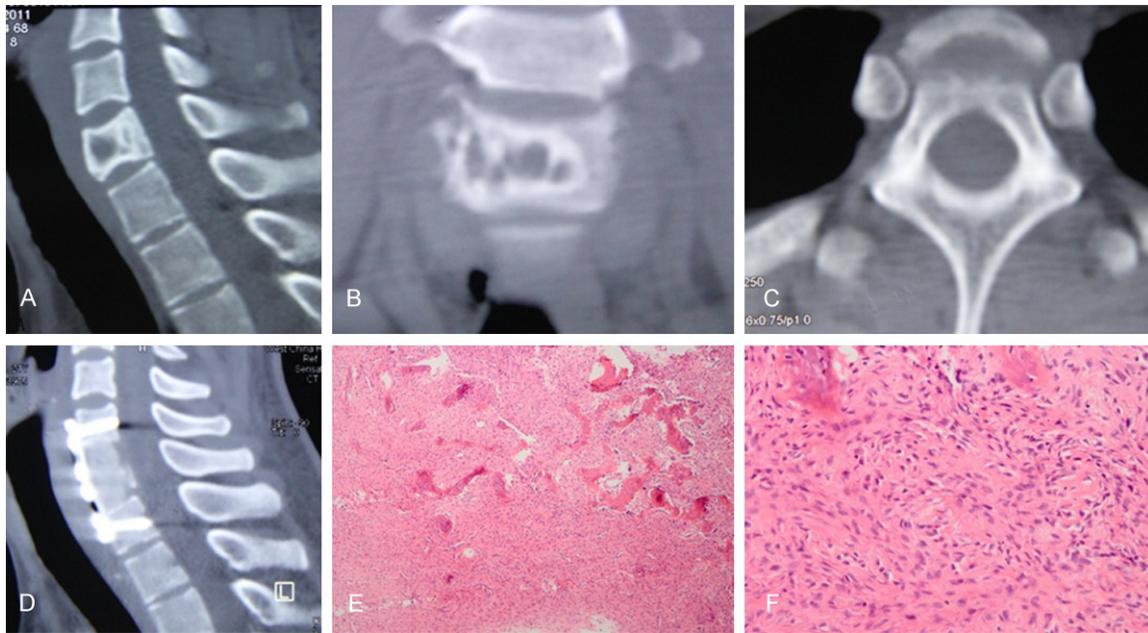


Figure 1. Radiographic and pathological images of case 1. A: Preoperative sagittal CT scan; B: Preoperative coronal CT scan; C: Preoperative axial CT scan; D: Postoperative sagittal CT scan; E: Pathological HE staining $\times 100$; F: Pathological HE staining $\times 400$.

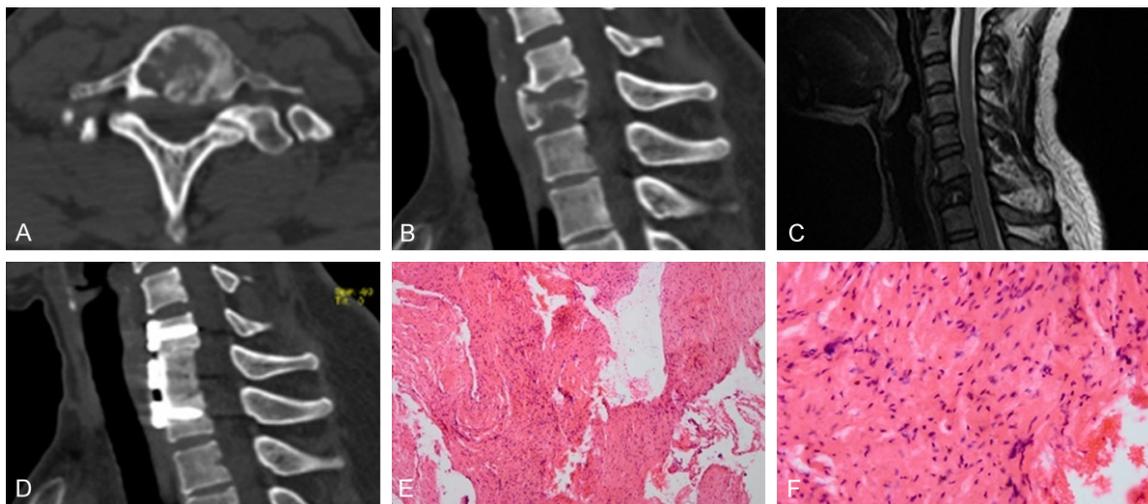


Figure 2. Radiographic and pathological images of case 2. A: Preoperative axial CT scan; B: Preoperative sagittal CT scan; C: Preoperative sagittal MRI; D: Postoperative sagittal CT scan; E: Pathological HE staining $\times 100$; F: Pathological HE staining $\times 400$.

showed bone destruction of the seventh cervical vertebrae and pedicles. Based on the lack of a past history of tumors, no evidence of other tumors from the findings of the systemic examinations, a preliminary diagnosis of primary spinal tumor was made. A surgery of anterior surgical resection, autogenous iliac bone graft with instrumentation of anterior plate was performed. Tissue surgically resected was sent to

department of pathology for pathological examination. Pathological hematoxylin and eosin (HE) staining results supported the diagnosis of fibrous dysplasia. Combined with the clinical examinations and radiographic results, the patient was finally diagnosed as MFD. Neither clinical symptoms nor signs of tumor recurrence were detected with a follow-up of more than 36 months. The radiographic and patho-

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Table 1. The characteristics of monostotic fibrous dysplasia (MFD) involved in cervical vertebrae*

NO.	Author	Year	Age	Gender	Presenting symptoms	Site	Initial treatment	Outcome
1	Schlumberger	1946	20	M	Posttraumatic	C4 (body)	Biopsy/observation	N/A
2	Rosendahl-Jensen	1956	35	F	Posttraumatic	C4 (body, lateral mass, posterior elements)	Curettage and bone graft	Well at 1-year follow-up
3	Resnik and Lininger	1984	27	F	Posttraumatic	C6 (body, lateral mass, lamina)	Biopsy/observation	N/A
4	Stirrat et al.	1989	25	M	Neck pain	C2 (body, odontoid)	Posterior fusion of occiput to C4	Asymptomatic at 2-year follow-up
5	Villas and Martínez-Peric	1992	11	M	Painful torticollis	C4 (body, L lateral mass)	Complete removal with A/P instrumented fusion	Asymptomatic at 4-year follow-up; no evidence of recurrence
6	Marshman et al.	2004	35	M	Pathologic fracture	C3 (body, R lateral mass)	Corpectomy with instrumented fusion	Asymptomatic at 18-month follow-up; no evidence of residual fibrous dysplasia invading the strut graft
7	Proschek et al.	2007	56	M	Neck pain	C4 (body, posterior elements)	Biopsy/observation	Minor pain and no increase in lesion size at 6-month follow-up
8	Schoenfeld et al.	2010	39	M	Incidental finding	C7 (body, spinous process)	Biopsy/observation	Asymptomatic at 3-year follow-up; no change in lesion size
9	Kotil and Ozyuvaci	2010	55	M	Neck pain	C2 (entire vertebra)	Vertebroplasty	Remarkable improvement in clinical relief of neck pain at 1-year follow-up
10	Meredith et al.	2011	41	M	Neck pain	C2 (entire vertebra)	C1-C3 posterior fusion	Well at 20-year follow-up with evidence of graft invasion by fibrous dysplasia
11	Zhong et al.	2015	38	F	Posttraumatic	C7 (body, posterior elements)	Biopsy/observation	N/A: the patient refused surgery on account to indulge herself in superstition at last.
12	Yang et al. (Case 1 in this study)	2016	21	M	Neck pain	C7 (body)	Corpectomy with instrumented fusion	Well at 36 months follow-up duration
13	Yang et al. (Case 2 in this study)	2016	42	F	Neck pain	C7 (body)	Corpectomy with instrumented fusion	Well at six months follow-up duration

*F, female; M, male; N/A, not available; A/P, a combined anterior and posterior fusion procedure.

logical images of this patient were summarized in **Figure 1**.

Case 2

A 42-year-old female patient was admitted to our hospital with a history of neck pain for 3 years. She had no symptoms of numbness, dizziness, weakness, and abnormal sensation. No obvious abnormality was detected through neurological and physical examinations. Laboratory findings were all within normal limits. Cervical X-rays, CT scan and MRI showed the bone destruction of the seventh cervical vertebrae and pedicles. No evidence of other tumors or lesions was detected through the systemic examinations. Anterior surgical resection, autogenous iliac bone graft with instrumentation of anterior plate was performed after general anesthesia. Grossly, the tumor was grayish-white and elastic hard. Pathological HE staining results supported the diagnosis of fibrous dysplasia. Based on the lack of a past history of tumors, no evidence of other tumors from the findings of the systemic examinations, the patient was finally diagnosed as MFD. With six months follow-up duration the patient had no clinical symptoms and no signs of tumor recurrence was detected. Her radiographic and pathological images were summarized in **Figure 2**.

Discussion

A sporadic activating mutation of Gs alpha on chromosome 20q13.2-13.3 has been reported to be associated with the occurrence of FD but the mechanism of FD has not been fully understood at present [3]. In MFD patients the mutated cells are thought to be limited to the affected bone but additional areas can also be affected (fail to establish clinical disease because of the competitive growth disadvantage compared with the non-mutated cells and increased rates of apoptosis of the mutated cells). In our literature review, 11 cases of MFD involved in cervical vertebrae were identified and characteristics of these cases (including two cases in this study) were summarized in **Table 1** [4-14]. There were 4 female patients and 9 male patients with an average age of 34.2 years. The presenting symptoms were neck pain in six patients, posttraumatic in 4 patients, pathologic fracture in one patient, painful torticollis in one patient, and incidental finding in one patient. The treatment method included: biopsy/observation in 5 patients, cor-

pectomy with instrumented fusion in 3 patients, posterior fusion in two patients, vertebroplasty in one patient, curettage and bone graft in one patient, and complete removal with a combined anterior and posterior fusion procedure in one patient. Outcomes of these patients have been uniformly good and the mean follow-up time is 3.65 years with a range of six months to 20 years while three patients' outcomes were not available.

Clinical and radiological examinations are not specific but necessary to exclude other diseases such as metastatic tumors because histological examination cannot define the origin of the tissue. Main differential diagnosis includes metastatic lesions, bone cysts, enchondroma, histiocytic fibroma and malignant primary bone tumours. Histopathological examination is critical for the diagnosis of FD. Stroma shows fibrous tissue with plump fibroblasts and formations of osteoid substance not assuming the aspects of osteoblasts. Bone trabeculae are presented in a uniform and woven structure with no atypic cells or mitotic hyperactivity. Whether, when and how to treat a FD involved in cervical vertebrae still remains controversial. A non-operative treatment such as observation can be followed as long as there are only minor complaints, no damage of cervical stability and no compromise of neurological structures. Surgical intervention is indicated if there is a mechanical reason for the pain or a cervical instability or even a compromise of neurological structures. Different kinds of surgeries can be effective such as arthrodesis, laminectomy, corpectomy and local excisions. There is no gold standard in operative treatment method and each treatment should be individualized to relief the symptoms, reconstruct cervical stability and avoid compromise of neurological structures.

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Disclosure of conflict of interest

None.

Abbreviations

FD, fibrous dysplasia; MFD, monostotic fibrous dysplasia; PFD, polyostotic fibrous dysplasia;

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CT, computed tomography; MRI, magnetic resonance imaging; HE, hematoxylin and eosin.

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