Int J Clin Exp Med 2015;8(11):21813-21817
www.ijcem.com /ISSN:1940-5901/IJCEM0014951

Case Report

Tumors and pseudotumors at the temporomandibular joint region in pediatric patients

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Received August 23, 2015; Accepted October 27, 2015; Epub November 15, 2015; Published November 30, 2015

Abstract: Objective: To describe the clinical manifestations and types of, and our surgical experience with, neoplasms in the region of the temporomandibular joint (TMJ) in pediatric patients. Materials and Methods: From September 1997 to December 2013, a total of 18 patients with neoplasms in the region of the TMJ were treated at our department. They all underwent open surgeries. The clinical manifestations and radiological aspects of all the patients were reviewed. The average follow-up period was 61.8 months with a range of 12-221 months. We reviewed the history, physical examination, images, and related radiological examinations. Results: Of the 18 patients, 14 had benign tumors or pseudotumors, and four had malignant tumors. The ratio of pseudotumor to benign tumor to malignant tumor was 2.5:1:1. Limitations of mouth opening were more likely to occur with malignant tumors, and facial deformity had a higher incidence in benign tumors. Local resection was the first choice for patients with benign tumors or pseudotumors. All patients with malignant tumors underwent whole-tumor resection along the boundary, including the joint capsule, disc, and part of the temporal bone and mandible. During the follow-up period, no tumor reformation or new deformity was detected. Conclusions: In the diagnosis of masses in the TMJ region, CT and MRI play an important role. Surgical removal of the mass with/without joint attachment was sufficient to treat benign and malignant tumors.

Keywords: Pediatric patients, temporomandibular joint, tumor, pseudotumor, surgery

Introduction

The temporomandibular joint (TMJ) is composed of the temporal bone, glenoid fossa, articular tubercle, mandibular condyle, articular disc, the articular capsule, and related ligaments [1, 2]. Tumors and pseudotumors in the mandibular joint area refer to the occurrence of neoplasms in the component parts of the temporomandibular joint. However, TMJ pseudotumors and tumors are infrequent, compared with the incidence of TMJ internal disorder (ID), which is as high as 28-88% among teenagers [3, 4]. However, there may be no significant difference in the clinical manifestations among tumors, tumor-like lesions, and ID. Thus, it is easy to misdiagnose and delay treatment [3], causing increased suffering, a greater risk of treatment complications, and, in the case of malignant tumors, increased threat to patient survival. We reviewed 18 patients under 18 years old with tumors and tumor-like lesions in the TMJ area from September 1997 to December 2013 in our department. We sought to summarize the category, clinical characteristics, imaging features, and prognosis for this type of disease.

Materials and methods

The ethics committee of Shanghai Jiao Tong University School of Medicine Affiliated No. 9 People's Hospital approved this study. We also obtained written informed consent from the parents on behalf of their children.

From September 1997 to December 2013, there were 18 patients under 18 years old diagnosed with tumors or pseudotumor lesions in the TMJ histologically at the Oral and Maxillofacial Surgery Dept., Shanghai Ninth People's Hospital, Shanghai Jiaotong University

Table 1. Types of pseudotumors and tumors

Types	Case
Pseudotumors (10)	
Osteofibroso heterohyperdysplasia	3
Langerhans cell histiocytosis	2
Condylar hyperplasia	1
Condylar cyst	3
Inflammatory lesions	1
Benign tumor (4)	
Osteoblastoma	2
Ossifying fibroma	1
Osteochondroma	1
Malignant (4)	
Synoviosarcoma	1
Myelocytoma	1
Aggressive fibromatosis	2

Table 2. Distribution of the TMJ masses

Types / Age (y)	Pseudotumor (n, %)	Benign tumor (n, %)	Malignant tumor (n, %)	Total (n, %)
0-6	2, 11.1	0, 0	2, 11.1	4, 22.2
6-12	3, 16.7	0, 0	0, 0	3, 16.7
12-18	5, 27.8	4, 22.2	2, 11.1	11, 61.1

School of Medicine. Neoplasms of the mandibular coronoid process were not included in the study. We reviewed the age, gender, symptoms and signs, radiological examinations, operation records, and pathologic diagnosis of the patients. Follow-up visits were performed for all patients.

Results

Category

Of the 18 patients, there were 10 (55.6%) with pseudotumor lesions, 4 (22.2%) with benign tumors, and 4 (22.2%) with malignant tumors (**Table 1**). If the pseudotumor lesions were grouped with the benign tumors, then the benign to malignant tumor ratio was 3.5:1. The pseudotumor lesion to tumor ratio was 2.5:1. The pseudotumor lesions included osteofibroso heterohyperdysplasia (3/10), condylar cysts (3/10), Langerhans cell histiocytosis (2/10), condylar hyperplasia (1/10), and an inflammatory lesion (1/10). Benign tumors included osteoblastoma (2/4), ossifying fibroma (1/4), and osteochondroma (1/4). The malignant tumors included aggressive fibromatosis (2/4), synoviosarcoma (1/4), and myelocytoma (1/4).

Age and sex distribution

Nine male and nine females patients were included (1:1). Five males and five females were ultimately diagnosed with pseudotumor lesions. Three male patients and one female patient had benign tumors. One male patient and three female patients had malignant tumors. The average age of the patients was 12.3 (range, 3-18) years; 4 (22.2%) cases were 6 years or younger, 3 (16.7%) were 6-12 years, and 11 (61.1%) cases were older than 12 years (**Table 2**).

Clinical and imaging findings

There were eight cases on the left and 10 on the right (left:right = 1:1.25). In 9 (50%) cases, there were limitations in mouth opening (less than 30 mm), 2 (11.1%) with snapping jaw, 6 (33.3%) with pain, 8 (44.4%) with malocclusion, and 10 (55.6%) with facial deformities. Additionally, in 2 (16.7%) cases, we palpated lymphadenovarix at the neck in the synoviosarcoma. One case with myelocytoma felt slight numbness in the affected lower lip. The incidences of facial deformity in pseudotumor lesions, benign tumors, and malignant tumors were 40%, 100%, and 50%, respectively. Limitations in mouth opening in the tumors, benign tumors, and malignant tumors were seen in 60%, 0, and 75%, respectively. Other clinical manifestations showed no obvious differences (**Table 3**). Langerhans cell histiocytosis, diagnosed by pathology, was isolated and did not occur with disease in other bones or organs CT or MRI scans were taken in all patients. Clear occupying lesions were revealed. For benign lesions, we found preserved structure of the condyle with well-margined and nearly homogeneous density (**Figure 1A** and **1B**). For malignant tumors, the condyle was replaced by the lesion completely, the edge was unclear, and the density was non-homogeneous on CT (**Figure 2A**) and MRI (**Figure 2B**). Approximately 40% of the pseudotumors, 50% of the benign tumors, and 25% of the malignant tumors showed osteogenic changes on CT. There was no statistically significant difference. Condylar absorption performance was 70%, 50%, and 100% in the

Table 3. Clinical features and imaging characteristics of TMJ tumors and pseudotumors

Appearance	Types	Pseudotumor lesion	Benign tumor	Malignant tumor
Clinical manifestation	snapping jaw	0	2	0
	limitation in mouth opening	6	0	3
	pain	3	2	1
	numbness in lower lip	0	0	1
	malocclusion	4	2	2
	lymphadenovarix	0	0	2
	facial deformity	4	4	2
Image	disc replacement	0	1	0
	perforation	0	1	0
	osteosclerosis	1	1	0
	condyle hyperplasia	4	2	1
	condyle absorption	7	2	4
	destruction of the base of the skull	3	0	2
Recurrence		0	0	0

pseudotumor, benign, and malignant lesions, respectively (P = 0.328). Destruction of the skull base was found in 30% of the pseudotumors, none of the benign tumors, and 50% of the malignant tumors (P = 0.328). Thus, condylar osteogenesis, condylar absorption, and destruction of skull base were not specific characteristic of these three lesions.

Surgical methods and follow-up results

All patients underwent surgical operations. For the benign lesions, local resection of the lesion was performed in 8 of 14 patients and a total condylar resection in 6 of 14 patients, due to more than 2/3 of the condylar surface being involved. For the malignant tumors, whole-tumor resection along the boundary with the capsule, disc, a portion of the temporal bone, and part of the mandibular ramus was performed. The same principles of reconstruction were applied to both benign and malignant tumors. A costochondral graft (CCG) was used in six cases to reconstruct the condyle. A temporal fascial fat flap (TFFF) was used to repair the small perforation of the skull base in one case of myelocytoma and one case of Langerhans cell histiocytosis. A TFFF and an autogenous non-vascular free ilium graft were also needed to reconstruct a large defect in the skull base in one case of Langerhans cell histiocytosis. Orthognathic surgery was also performed due to a severe maxillofacial deformity. The mean follow-up period was 61.8 (range, 3-221) months. After the operation, pain, swelling, and limitations on mouth opening were resolved. Also, facial deformity and malocclusion were corrected. No case of Frey syndrome or permanent facial paralysis occurred. No postoperative recurrence was found in any patient. Two CCGs showed overgrowth (2/6).

Discussion

Tumors and pseudotumor lesions rarely occur in the TMJ. According to the literature, in adults, benign tumors primarily include chondroblastomas, osteoblastomas, osteochondromas, and osteomas, while metastatic tumors and sarcomas are the main malignant tumors [5]. In our study of young patients, osteofibroso heterohyperdysplasia, condylar cyst, and Langerhans cell histiocytosis were the major pseudotumor lesions, osteoblastoma was the main benign lesion, and aggressive fibromatosis was the main malignant tumor. In a past report, we found that the ratio of pseudotumor to benign tumor to malignant tumor in adults was 7.5:1:2 [5], which differed from our finding of a 5:2:2 ratio in adolescents. For adults, the incidence in females was significantly higher than that in males [6] for pseudotumor lesions, benign tumors, and malignant tumors. In these 18 cases of pediatric patients, we found no significant gender difference in pseudotumor lesions, but there were more males with benign tumors and more females with malignant tumors.

Due to their similar clinical manifestation to that of TMJ disorder, it can be difficult to diagnose tumors directly [7]. However, we found that numbness of the lower lip and lymphadenovarix only occurred in the malignant tumor

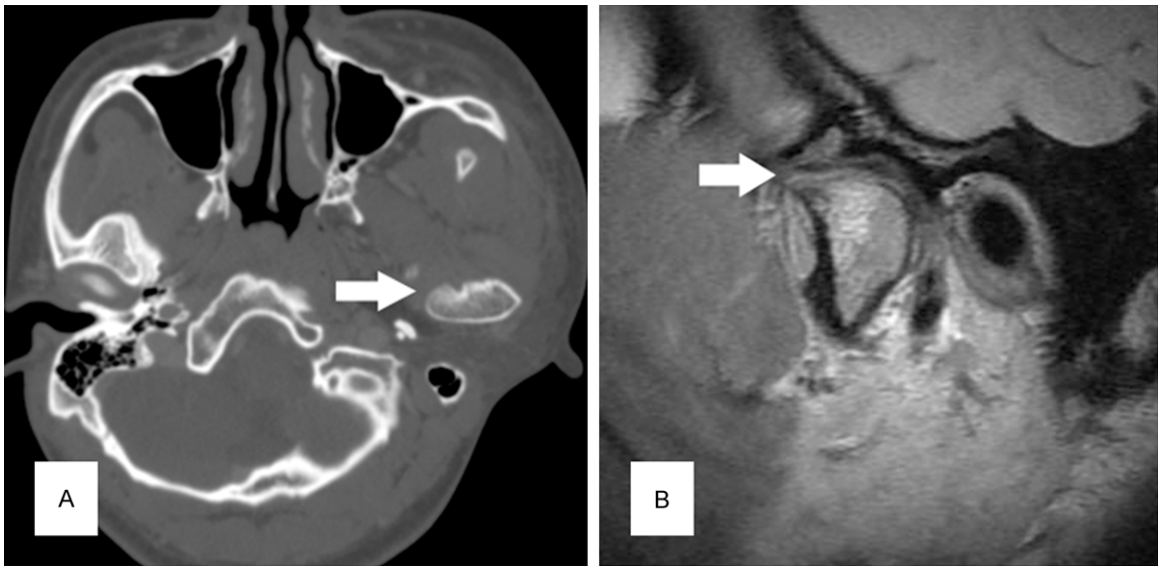

Figure 1. A. Condylar hyperplasia on CT. B. Condylar osteophyte on MRI.

Figure 2. A. Condylar destruction by aggressive fibromatosis on CT. B. Replacement by aggressive fibromatosis on MRI.

cases in this study, which are alert signals for malignant tumors. CT and MR were helpful in making the diagnosis of tumor or pseudotumor lesions. However, radiological images do not always distinguish pseudotumor lesions, benign tumors, and malignant tumors. In our study, there was no significant difference in condylar osteogenesis, condylar absorption, and destruction of the skull base among these three lesions. Thus, a pathological diagnosis was needed for the final diagnosis to clarify the type of mass.

According to intraoperative frozen pathological diagnoses, different methods were used. For benign lesions, local resection with preservation of the condyle as much as possible was the priority. For malignant tumors, radical resection along the boundary, including the disc and articular capsule, was necessary. To benefit the function of the TMJ and growth of the mandible, a CCG was considered to be the best choice for condyle reconstruction [8-10], especially for pediatric patients [11, 12]. However overgrowth of the CCG, which may lead to recurrence of maxillofacial deformity, deserves attention. Tumor resection could lead to a large dead space. TFFF [13-15], a free fat flap [16], or a buccal fat pad 14 was needed to fill the dead

space, reducing the risk of postoperative infection and preventing ankylosis [17, 18].

In conclusion, there was no specific clinical manifestation for identifying pseudotumor lesions, benign tumors, and malignant tumors. CT and MRI are helpful in distinguishing the mass from TMJ disorders effectively. However, sometimes, CT and MRI cannot distinguish benign from malignant lesions. The final diagnosis still depends on intraoperative frozen biopsies and postoperative histopathology. Surgery remains the preferred treatment.

Acknowledgements

We thank the Pathology and Imaging departments for their assistance. This project was supported by National Natural Science Foundation of China (No. 81100824), Foundation of Shanghai Municipal Education Commission (No. 12YZ044), Foundation of Shanghai Municipality Science and Technology Commission (No. 14411960800), Technology Commission of Shanghai Municipality Science Research Project (No. 14DZ2294300) and Medical engineering cross project of Shanghai Jiaotong University (No. YG2013MS59).

Disclosure of conflict of interest

None.

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References

[1] Alomar X, Medrano J, Cabratosa J, Clavero JA, Lorente M, Serra I, Monill JM, Salvador A. Anatomy of the temporomandibular joint. Semin Ultrasound CT MR 2007; 28:170-183.

[2] Speber GH. Temporomandibular joint. In: Speber GH, editor. Craniofacial development. Hamilton, Canada: BC Deker; 2001. pp. 139-14.

[3] Guralnick W, Kaban LB, Merrill RG. Temporomandibular joint afflictions. N Engl J Med 1978; 299: 123-129.

[4] Ferendiuk E, Zajdel K, Pihut M. Pihut Incidence of Otolaryngological Symptoms in Patients with Temporomandibular Joint Dysfunctions. Biomed Res Int 2014; 2014: 824684.

[5] Poveda-Roda R, Bagán JV, Sanchis JM, Margaix M. Pseudotumors and tumors of the temporomandibular joint. A review. Med Oral Patol Oral Cir Bucal 2013; 18: e392-402.

[6] Poveda-Roda R, Bagán JV, Jiménez-Soriano Y, Fons-Font A. Retrospective study of a series of 850 patients with temporomandibular dysfunction (TMD). Clinical and radiological findings. Med Oral Patol Oral Cir Bucal 2009; 14: e628-34.

[7] Allias-Montmayeur F, Durroux R, Dodart L, Combelles R. Tumours and pseudotumorous lesions of the temporomandibular joint: a diagnostic challenge. J Laryngol Otol 1997; 111: 776-81.

[8] Kaban LB, Perrott DH, Fisher K. A protocol for management of temporo- mandibular joint ankylosis. J Oral Max- illofac Surg 1990; 48: 1145-1151.

[9] Matsuura H, Miyamoto H, Kurita K, Goss AN. The effect of autogenous cost- ochondral grafts on temporomandibular joint fibrous and bony ankylosis: A pre- liminary experimental study. J Oral Max- illofac Surg 2006; 64: 1517-1525.

[10] Ware WH, Brown GL. Growth centers transplantation to replace mandibular condyles. J Oral Maxillofacial Surg 1981; 9: 50-58.

[11] El-Sayed K M. Temporomandibular joint reconstruction with costochondral graft using modified approach. Int J Oral Maxillofac Surg 2008; 37: 897-902.

[12] Kaban LB, Bouchard C, Troulis MJ. A Protocol for Management of Temporomandibular Joint Ankylosis in Children. J Oral Maxillofac Surg 2009;67: 1966-1978.

[13] Smith JA, Sandler NA, Ozaki WH, Braun TW. Subjective and objective assessment of the temporalis myofascial flap in previously operated temporomandibular joints. J Oral Maxillofac Surg 1999; 57: 1058-1065.

[14] Bergey DA, Braun TW. The posterior zygomatic arch osteotomy to facilitate temporalis flap placement. J Oral Maxillofac Surg 1994; 52: 426-427.

[15] Omura S, Fujita K. Modification of the temporalis muscle and fascia flap for the management of ankylosis of the temporomandibular joint. J Oral Maxillofac Surg 1996; 54: 794-795.

[16] Candirli C, Esen A, Taskesen F, Celik S, Cakir B. Evaluation of discectomyand dermis-fat grafting on the contralateral side. J Craniofac Surg 2012; 23: 1654-1657.

[17] Dimitroulis G, Slavin J, Morrison W. Histological fate ofabdominal dermis-fat grafts implanted in the temporomandibularjoint of the rabbit following condylectomy. Int J OralMaxillofac Surg 2011; 40: 177-183.

[18] Dimitroulis G, Trost N, Morrison W. The radiological fate of dermisfat grafts in the human temporomandibular joint usingmagnetic resonance imaging. Int J Oral Maxillofac Surg 2008; 37: 249-254.