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Original Article

Primary intra- and juxta-articular vascular malformations of the temporomandibular joint: a clinical analysis of 8 consecutive patients

Qin Zhou, Chi Yang, Min-Jie Chen, Ya-Ting Qiu, Wei-Liu Qiu, Jia-Wei Zheng

Department of Oral and Maxillofacial Surgery, Ninth People's Hospital, College of Stomatology, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China

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Abstract: Objective: To analyze primary intra- and juxta-articular vascular malformations of the temporomandibular joint. Patients and methods: This study retrospectively reviewed eight patients (seven venous malformations and one lymphatico-venous malformation) who were treated for intra- or juxta-articular vascular malformations of the temporomandibular joint from November 2005 to January 2011. All patients underwent magnetic resonance imaging (MRI) preoperatively. Results: According to MRI findings, vascular malformations involving TMJ could be divided into 3 types; homogenous, lacunar and mixed types. All patients underwent surgical resection, and the final clinical diagnoses were confirmed by postoperative histopathology and immunohistochemical examinations. All treated patients had no clinical or radiographic signs of recurrence. Conclusion: Owing to the lower incidence and nonspecific clinical presentations, preoperative diagnosis of vascular malformations involving the TMJ region is very difficult. The classification based on MRI manifestations is proposed first, then it may greatly help in the initial diagnosis. Surgical resection is considered the first option for these TMJ lesions with excellent results.

Keywords: Vascular malformations, venous malformation, temporomandibular joint (TMJ)

Introduction

Vascular anomalies are congenital aberrations of vascular development. Based on their endothelial properties, they can be divided into two main categories: hemangiomas and vascular malformations [1]. Hemangiomas are the most common benign vascular tumors with abnormal endothelial proliferation, characterized by excessive proliferation in the first year of life followed by spontaneous regression thereafter. While vascular malformations are collections of enlarging aberrant and ectatic vessels, displaying normal flat non-proliferative endothelium, which are present at birth, growing commensurate with the patient throughout life and never involute. The proper diagnosis of vascular anomalies is extremely challenging because of the inconsistent terminology and classification systems. Hassanein et al [2] reported that "hemangioma", as a terminology, was used incorrectly in 71.3 percent of publications to describe any type of vascular anomaly. Vascular malformations can be further subdivided into low-flow (capillary, venous, lymphatic or combinations) and high-flow (arterial component, typically arteriovenous) lesions based on blood flow dynamics and basic type of the predominant constituent vessel [3].

Although any region of the body may be affected, the head and neck area is the most common site to be involved. Clinically, low-flow vascular malformations usually manifest as typically soft, compressible masses, and in case of venous malformations, lesions can enlarge with dependent positions and Valsalva maneuver [4]. High-flow lesions, like arteriovenous malformation (AVM), are rare and usually present with hyperemia, increased temperature, pulsatility, and palpable local thrill. Vascular malformations in the head and neck region commonly occur in the mucosal or cutaneous surfaces having a variable blue-tinged appearance. However, deep lesions are often associated with delayed presentation due to lack of obvi-

Table 1. Characteristics of the patients in the study

Case	Age/Sex	Joint right/left	Trauma yes/no	Symptoms	Duration of Symptoms (months)	Imaging examination	Follow up (months)
1	69/F	R	NO	Pain	24	MRI	114.5
2	44/F	L	NO	Pain, LMO	2	CT, MRI	87
3	24/F	L	NO	Pain, LMO, Swelling	1	CT, MRI	50
4	53/F	L	NO	Swelling	120	MRI	52
5	72/F	L	NO	Pain, LMO	5	MRI	45
6	64/M	R	NO	Clicking, Pain	4	CT, MRI	45.5
7	58/F	R	NO	Clicking, Swelling	24	CT, MRI	46
8	44/F	L	NO	Pain, LMO	36	CT, MRI	46

LMO: limitation of mouth opening.

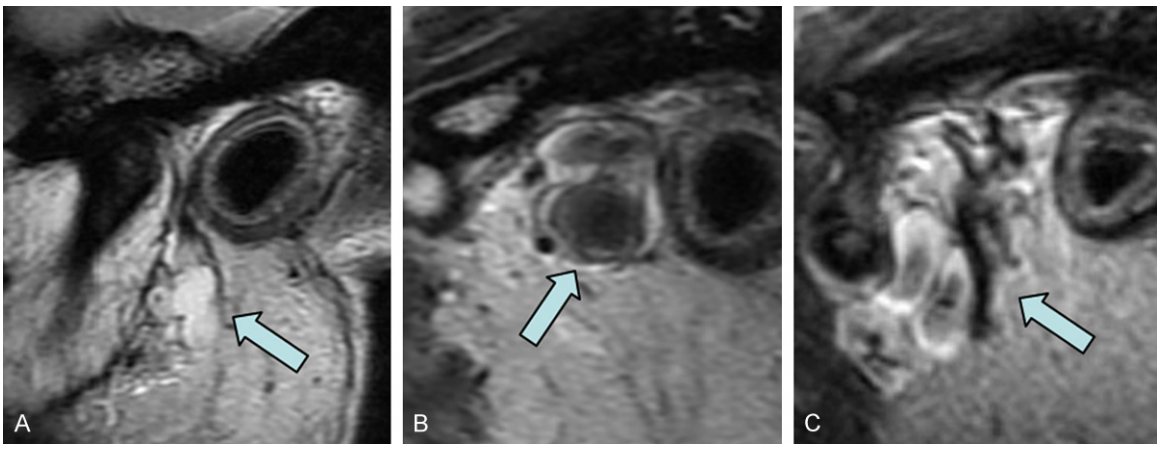

Figure 1. T2-weighted MRI image of intra- or juxta-articular vascular malformations of temporomandibular joint. A. Homogenous type, T2-weighted image shows hyperintense venous malformation (arrow) near the left TMJ with rounded foci of lower signal within the lesion likely representing phleboliths. B. Lacunar type, T2-weighted image shows central low signal with peripheral high density (arrow). C. Mixed type, homogenous and lacunar findings were found simultaneously (arrow).

ous skin involvement or deformity until they attain a certain size [5, 6]. Radiographic investigations including ultrasonography, plain radiography, CT scanning, and MRI aid in the diagnosis. When the characteristic clinical and radiological features are evident, diagnosis is not difficult. Lesions involving the temporomandibular joint (TMJ) regions usually lead to cosmetic and functional problems resembling parotid tumors or disorders of the TMJ. So the primary diagnosis of vascular lesions is extremely rare, and the definitive diagnosis is commonly confirmed after histological examination.

As a result of the low incidence of such anomalies in the region of the TMJ, most literature descriptions of vascular lesions in this area are case reports [7-9], therefore, no large series of cases have been published so far. The aim of this study is to provide information on clinical features, radiographic and histological characteristics of eight patients who were treated for an intra- or juxta-articular vascular malformations of the TMJ in the authors' department.

Materials and methods

Eight patients diagnosed as intra- or juxta-articular vascular malformations of the TMJ, with surgical resection of the lesions in the TMJ division of Shanghai's Ninth People's Hospital (Shanghai, China) from November 2005 to January 2011, were retrospectively reviewed. All the pathological slides and immunohistochemical assessment of the eight patients were re-examined carefully. Patients were referred for consultation with the chief com-

Table 2. Preoperative and postoperative diagnosis of the patients in this study

Case	Preoperative diagnosis	Postoperative diagnosis
1	SC or AVM	VM
2	Preauricular neoplasm	VM
3	Tumor of deep parotid or condyle	VM
4	mesenchymal or adenogenous tumors	VM
5	SC	VM
6	Giant cell granuloma	LM-VM
7	adenogenous tumors	VM
8	Malignant tumor	VM

SC: synovial chondromatosis; AVM: arteriovenous malformations; VM: venous malformations; LM: lymphatic malformations.

plaints of pre-auricular pain, impaired mandibular function, and swelling in the TMJ area. The information regarding their age, sex, site, symptoms, duration of symptoms, imaging examination and follow up are shown in **Table 1**. All patients underwent open surgery to the TMJ and lesions were successfully resected.

Results

There were 7 females and 1 male, with an average age of 53.5 years (range, 24-72 years) at the time of surgery. The average duration to first visit was 27 months (range, 1-120 months). The predominant complaints were limitation of mouth opening and pre-auricular pain or swelling. The maximal inter-incisal opening varied from 1.0 to 4.1 cm (average 2.6 cm). Pre-auricular swelling was found in three patients, pre-auricular pain and/or tenderness in six, and limitation of mouth opening in four. All patients had no history of trauma. Case 6 was complicated with chronic inflammatory granulation tissue in the right external auditory canal.

All patients underwent maxillofacial CT and/or MRI examination preoperatively. CT showed well or ill-defined soft-tissue masses, and in case 7, phlebolith was seen. Based on the findings from MR imaging, lesions were classified into three types: homogenous (4/8 patients), lacunar (3/8 patients) and mixed type (1/8 patients). Lesions with homogenous type were usually manifested as well-defined soft tissue masses with isointense signal on T1 and hyperintense on T2 (**Figure 1**). Lacunar lesions appeared to be of low signal in central areas and high signal in the periphery. Mixed lesions displayed the two forms simultaneously. The primary sites of the lesions were: anteromedial part of the condyle (1 patient), anterior part (2 patients), posteromedial part (1 patient), posterolateral part (3 patients), and in 1 patient, the lesion covered the condylar process from posteromedial to anterolateral part. The joint capsule was involved in 3 cases. In patients 1 and 2, vascular lesions were combined with right and bilateral non-reducing anterior disc displacement respectively, moreover, medial displacement of the right TMJ disc was seen in patient 8. On the basis of these findings, the initial diagnoses included benign tumors (synovial chondromatosis, adenogenous tumors, mesenchymal tumors, etc.) or low potential malignancy tumors of the parotid or TMJ (**Table 2**).

For all patients, intraoperative biopsy for frozen section was obtained. All masses were removed completely, with no significant hemorrhage during the procedure (The detailed operation steps will be addressed in a future article). Macroscopic examination of the resected specimens demonstrated dusky-red, firm, well circumscribed masses with an intact capsule. The section view showed numerous small sinuses filled with blood (**Figure 2**), but in case 7 nodular appearance was found, with flesh like texture (**Figure 3**). The final clinical diagnosis was confirmed by histological and immunohistochemical examination. The presented vascular malformation series comprised 1 lymphatico-venous malformation and 7 venous malformations. Microscopic examination revealed dilated or ectatic vascular channels lined by flat endothelium. Furthermore, these vascular spaces were usually filled with an abundance of erythrocytes, with more collagen fibrils found in four patients compared with that of typical vascular malformations (**Figure 4A**, **4B**). There were intermittent smooth muscle within the VM channel wall, in addition, lymphatic vessels filled with lymphatic fluid or much fewer erythrocytes could be seen in lymphatico-venous malformations. Immunohistochemistry is non-specific, as all patients were positive for CD31, CD34 and factor VIII (F8), and patients with venous malformations were positive for SMA in our series (**Figure 4C**, **4D**).

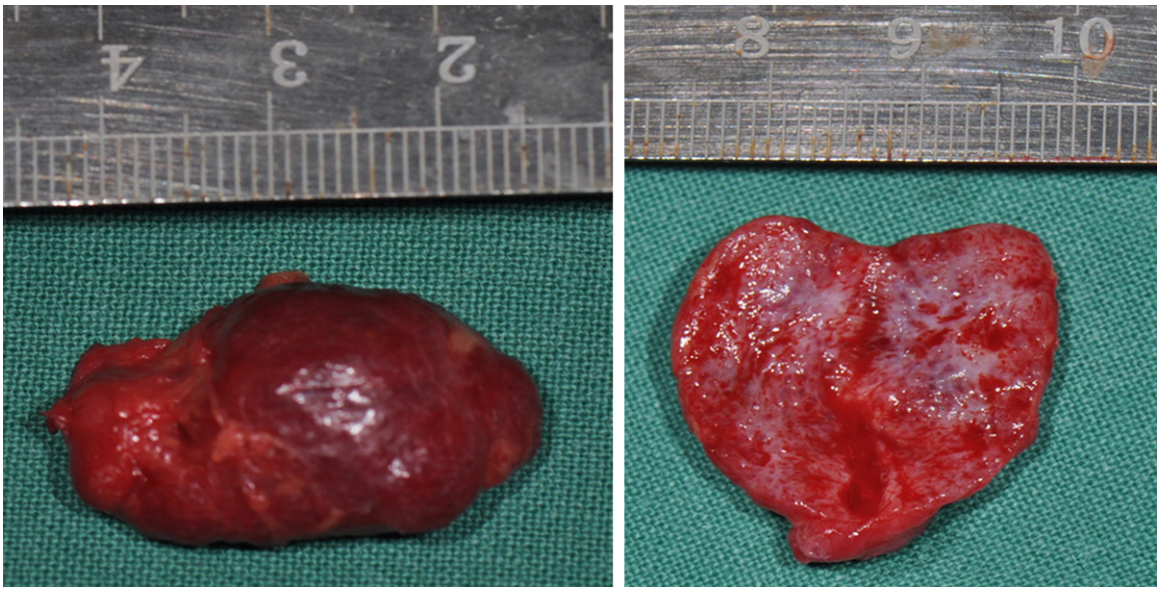

Figure 2. The dark red, soft surgical mass with intact thin capsule from patient no. 3, and the section with numerous small sinuses filled with blood.

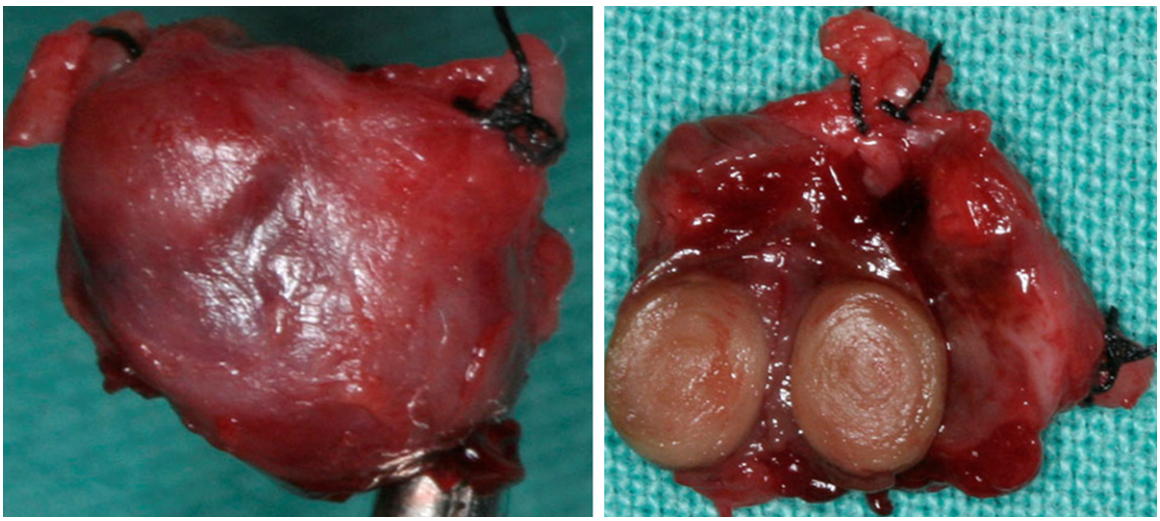

Figure 3. The dark red, soft surgical mass with intact thin capsule from patient no. 7, and the section texture like flesh.

Discussion

Venous malformations are the most frequent low-flow vascular malformations, the majority are sporadic and unifocal, about 1% are multifocal [10]. Lesions are present since birth but not always evident clinically till later. Little is understood regarding the pathogenesis. Trauma or infection is all known to result in increased growth or pain. In addition, hormonal influences, such as pregnancy or oral contraceptives, can also influence growth, although the exact mechanisms are not entirely clear [11]. In this series, seven cases were diagnosed as venous malformations and one as lymphatico-venous malformation, but they did not present with any obvious symptoms until recently. Common cold exacerbated the disease in case 2, and case 6 was complicated with chronic inflammatory granulation tissue in right external auditory canal.

Diagnosis is principally clinical and supported by imaging investigations. Venous malforma-

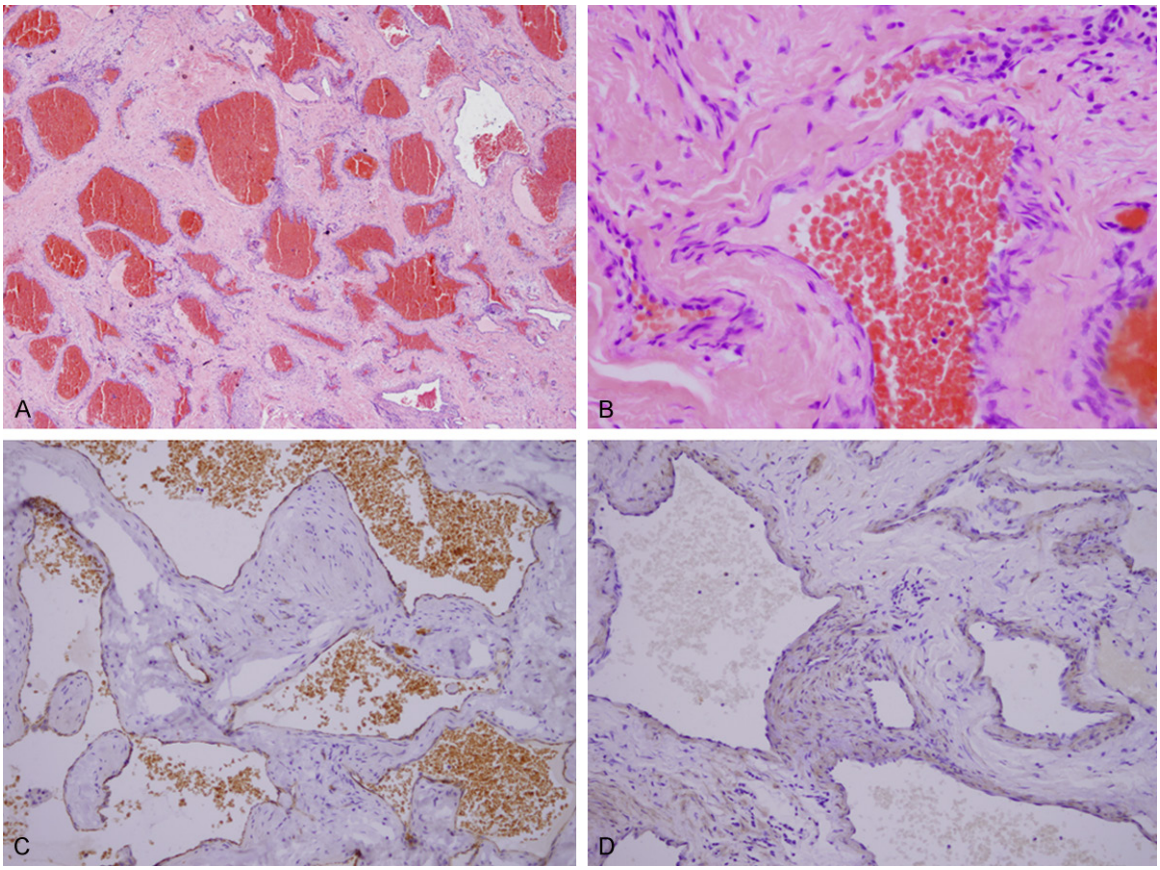

Figure 4. Histological examination of venous malformation from patient no. 3. A. Variably sized thin-walled veins filled with thrombosis (HE×40). B. Dilated venous channels lined by flattened endothelium, and rich collagen fibrils within mesenchyma (HE×400). C. Positive reactivity for CD31 was observed in the endothelium (×200). D. Positive reactivity for SMA was observed in the pericytes and smooth muscle cells (×200).

tions usually present clinically as soft, compressible blue masses that enlarge when the affected area is in a dependent position or with physical activity. Palpation is not painful unless thrombosis occurs. In seven patients in our series, the initial diagnosis was erroneous which highlighted the common delay in diagnosing vascular lesions involving the TMJ. Clinically, patients presented with no symptoms or nonspecific complaints. Because of the deep location, the size of the lesion does not alter with postural changes, in contrast to VMs in other parts of the head and neck region. Fine-needle aspiration cytology (FNAC) is also not easy to perform practically. Furthermore, lesions were usually not associated with a history of bleeding. The lack of clear objective clinical findings, and the rare incidence in this location, often lead to misdiagnosis. As a result of the complex structural anatomy in the pre-auricular region, the diagnosis of a mass in the area is much difficult, so many initial presumptive diagnoses must be considered.

The clinical differential diagnosis of intra- and juxta-articular vascular malformations of the temporomandibular joint should include synovial chondromatosis (SC), pigmented villonodular synovitis (PVNS), neurilemmoma and ganglion. Loose bodies of synovial chondromatosis on MRI are not so obvious but lesions just appear as a swelling in the lateral capsule, with associated fluid in the joint cavity. PVNS has variable appearances on MR images, as the hemosiderin pigments shorten both T1 and T2 relaxation time [12]. MRI is not helpful in the diagnosis of simultaneous PVNS and synovial chondromatosis, meanwhile, arthroscopy is a very reliable method. Ganglion cysts are pseudocysts with a fibrous connective tissue wall, thus lacking the synovial cell lining, and filled with viscoid or gelatinous material [13]. Sch-

wannoma is characterized by solitary, encapsulated mass, along the course of a nerve, and often complicated with symptoms of nerve irritation, which is dominated by spindle cells. They have nonspecific imaging findings, therefore, postoperative pathology can confirm the definitive diagnosis.

Imaging is necessary for diagnosis of indeterminate lesions, in addition, evaluating the extent of the lesion and associated abnormalities. The radiographic features of vascular anomalies are nonspecific, actually, they are known as "the great radiologic imitators", because the lesions can look like any lesion ranging from a cyst to a malignancy on radiograph [14]. Color Doppler ultrasound is usually used to evaluate the blood flow velocities and patterns. CT scanning is extremely sensitive in detecting phleboliths [15-17]. MRI is still the golden standard for diagnosis of vascular lesions. Venous malformations usually have an isointense or hypointense signals on T1 images with high signal intensity on T2 imaging [4, 18-20], lymphatic malformations often have low T1 and high T2 signal intensity, whereas a low signal on both T1 and T2 was seen in high-flow malformations, corresponding to the high-flow arteries and arterialized veins [21, 22]. Combined vascular malformations share the features of both low- and high-flow lesions. In this study, four of eight patients' MRI findings were homogenous type with isointense signal on T1 and hyperintense on T2. Three cases appeared as lacunar type and one as mixed type. So when these imaging characteristics are seen in the TMJ region, vascular lesions should be considered. In order to prevent misdiagnosis and missed diagnosis, we suggest that maxillofacial MRI should be obtained furthermore.

FNAC is impractical and extremely difficult owing to the deep position and the small sinus cavity of vascular malformations of the TMJ, meanwhile, histological examinations help to confirm the final diagnosis for surgical patients. Vascular malformations could be localized or extensive. Head and neck VMs are frequently more extensive than initially apparent, which may extend into adjacent regions resulting in cosmetic and functional problems. In this series, all lesions were firm, well circumscribed with a thin layer of intact capsule. In addition to the typical histopathological findings, much more collagen fibrils were also seen in 4 of 8 patients, which might be associated with small, well localized lesions. Immunohistochemical analysis is an important adjunctive diagnostic approach to establish the correct diagnosis for cases with mixed or ambiguous histology. CD31 is the most sensitive and specific of all antibodies for vascular endothelium [23]. D2-40 is a recently available monoclonal antibody that is reported to be a highly sensitive and specific marker for lymphatic endothelium [24].

Vascular malformations in the TMJ area are well localized, with abundant fibers and small sinus cavity. Therefore, sclerotherapy was considered an inappropriate modality and surgical treatment in our limited study sample resulted in good outcomes. The mean follow-up period was 60.75 months (range, 45 months to 114.5 months). During the follow-up period, symptoms were alleviated except in two patients with post-operative numbness. No recurrences were observed, and the quality of life of all patients was good.

In conclusion, although preoperative diagnosis of intra- or juxta-articular vascular anomalies of the TMJ is problematic, imaging classification in this series may help diagnosis in the future. Surgery should be considered as the first line of treatment in this area, having excellent results. As far as we know, this is the first large sample study on vascular malformations involving TMJ.

Disclosure of conflict of interest

None.

Address correspondence to: Chi Yang, Department of Oral and Maxillofacial Surgery, Ninth People's Hospital, College of Stomatology, Shanghai Jiao Tong University School of Medicine, NO. 639 Zhi Zao Ju Road, Shanghai 200011, China. Tel: +86-21-23271699-5218; Fax: +86-21-63135412; E-mail: yangchi63@hotmail.com

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