

*Full Length Research Paper*

# **Osteoma orbitalis: A case report and literature systematic review**

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**Orbital osteoma is a very rare benign tumor in orbit, and its diagnosis and treatment were certain particularity. Therefore, we reported one case of osteoma orbitalis and reviewed relevant literature, in order to further understand clinical features of osteoma orbitalis. A 38-year-old female was hospitalized due to left exophthalmos after frontal trauma for 5 years. Physical examination: left exophthalmos forward and laterally downward; a hard mass in the internal top of the eye sockets resulted in oppressive changes of ocular fundus. Movement of the left eye was slightly restricted. CT showed the bone-like mass while no findings were shown in MRI. Orbitotomy plus orbital tumor extirpation of the left eye were performed. Pathology indicated mature bone tissues, so osteoma orbitalis was considered. Systematic reviews of relevant literature showed that : the first case of osteoma orbitalis in China was reported in 1958, and a total of 40 cases have been reported up to now while a total of 47 cases have been reported since 1903 in Medline. Slightly higher incidence in females was found compared with males. The mean onset age was about 30 years old (27.04±10.8 years in China while 32 years old in foreign countries). The main clinical manifestations of osteoma orbitalis were as follows: exophthalmos (75%), fundus changes (60%), decreased vision (36.7%) and eye movement disorder (34%). Patient's systematic conditions and local non-orbital diseases were more overall reported in foreign literatures. The onset, clinical features and treatment of orbital tumors were special from other tumors, and the specificities should not be neglected.**

**Key words:** Osteoma orbitalis, case report, literature systematic review.

## **INTRODUCTION**

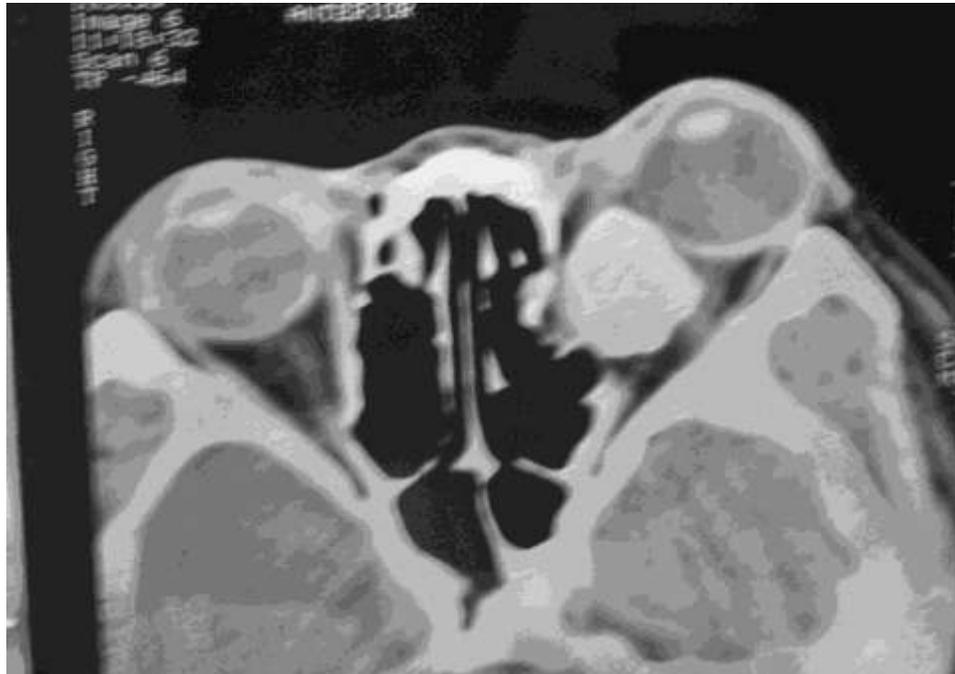
Osteoma orbitalis is a rare orbital space occupying disease. This disease has some distinguished clinical characteristic. For trying to find the clinical characteristic of osteoma orbitalis, a case was reported and Chinese clinical medicine literatures and some of papers from Medline were collected. Here, we found some differentiation which was caused by emphasis and standpoint in the two systems. This differentiation reflected there some distinction at clinical medicine in Chinese and West countries.

## **MATERIALS AND METHODS**

### **Case report**

A 38-year-old female was hospitalized on March 2, 2006 due to left exophthalmos after frontal trauma for 5 years. Physical examination: Vod: 1.0, Vos: 0.15, could not be corrected; left exophthalmos was forward and laterally downward; the degrees of the left and right exophthalmos were 22 and 14 mm, respectively. Movement of the left eye was slightly restricted. Palpation found a hard intraorbital mass in the internal top of the eye sockets, which adhered to internal and superior orbital wall, without activity or tenderness. Eyeball examination: no apparent abnormality was found in the anterior segment of the left eye, and the boundary of papilla optica was obscure, and varicose veins and retina folds with the center at the nasion were found in ocular fundus. CT revealed a

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**Figure 1.** CT revealed a high-density mass adhering to nasal sinuses.

large lesion in the left eye sockets and above the nose, and CT value was similar with the bone, without internal structure. There was obscure boundary between the ethmoid bone and the mass, and some mass tissues intruded into the ethmoidal sinus (Figures 1 and 2). No visualization was observed in same regions in MRI T1 or T2, only showing a cavity-like lesion in tissues (Figures 3 and 4). Color B-type ultrasonography showed a  $2.1 \times 2.65 \times 2.74$  cm irregular solid mass in the nasal side and behind the eyeball, with unclear borders. Enhanced superficial echo of the mass and rear echo attenuation were also observed (Figure 5), and the optic nerve was extruded to the temporal side, and the mass did not move with the movement of eyeballs. Clinical diagnosis of this case was left osteoma orbitalis.

Orbitotomy plus orbital tumor extirpation of the left eye were performed under general anesthesia. Intraoperative observation revealed that the intraorbital bony mass adhered to the orbital inner wall and some mass tissues intruded into the ethmoidal sinus. There was no obvious adhesion or blood supply between the tumor and surrounding tissues. The mass was too large to extract through the peri-eyeball space, and thus some superior border of orbital bone was removed. Grossly, it presented roughly oval and irregular tumor appearance, and the size of the tumor was  $2.4 \times 2.8 \times 3.2$  cm (Figures 6 and 7).

Eye examination at postoperative 7 days revealed 1.0 in Vos, 15 mm in exophthalmos, moderate left ptosis and slight restriction of ocular upward movement. No abnormality of eyeball or eye fundus was observed. Pathology revealed mature bone tissues, so osteoma orbitalis was considered (Figure 8).

## RESULTS

### Literature in Chinese

The first case of osteoma orbitalis in China was reported

by SHANG Chong-xue in 1958 (Shang, 1958). With the keywords of "Orbita" and "Osteoma", all medical literature in Chinese in VIP and Wan Fang databases from 1989 to 2008 were indexed on January 9, 2009. The results are listed as follows:

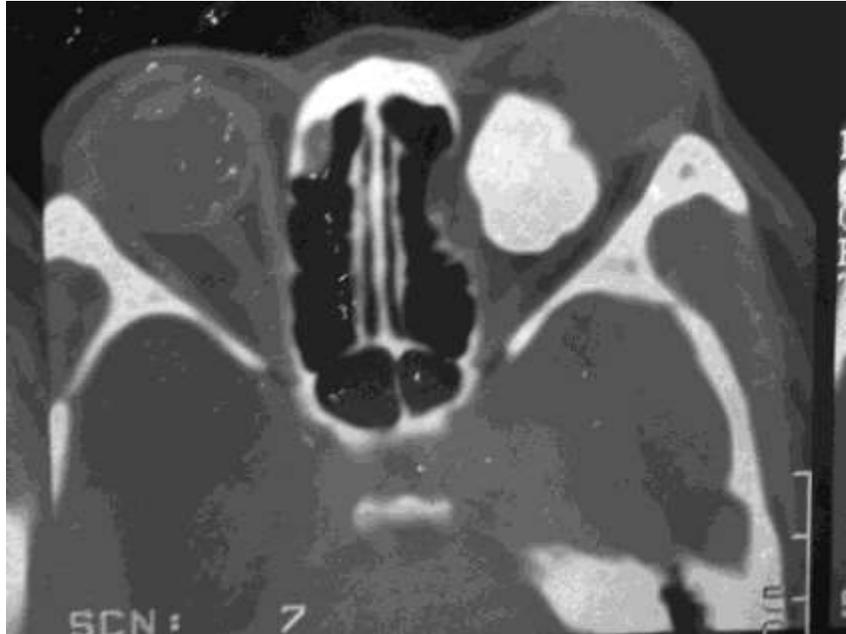
### VIP database

A total of 14 articles were primarily indexed. Based on the titles and abstracts, all articles about "choroidal osteoma" and "paranasal sinus osteoma accompanied with orbital lesions" were excluded, and thus only 9 articles met the requirements. Meanwhile, 18 articles were indexed in digital periodical link database, but none was selected after the removal of duplicate articles.

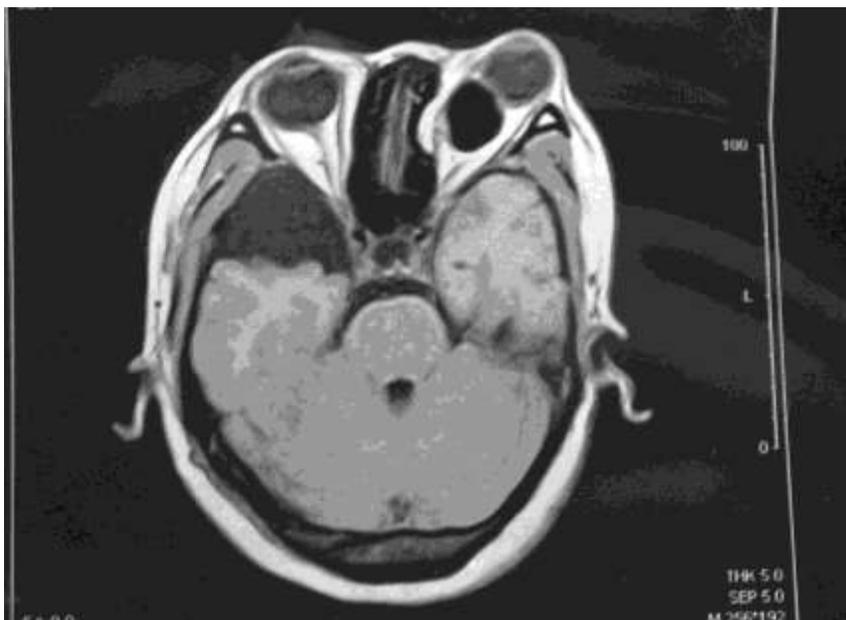
### Wan Fang database

A total of 17 articles were primarily indexed, but 8 articles were ruled out. Meanwhile, 9 articles were indexed in links and citation database, but none was selected after the removal of duplicate articles. Thus, 9 articles meet the requirements.

There were 5 duplicate articles in VIP and Wan Fang databases, and 13 articles were eventually indexed in databases. However, 7 articles were selected from the citations in the 13 indexed articles. Thus, a total of 20 articles in Chinese met the requirements, including 5 original articles and 15 case reports, with 59 cases of



**Figure 2.** CT enhancement scanning revealed the density of mass was uniform and similar with bones.



**Figure 3.** MRI (T1) revealed a cavity-like lesion in tissues.

osteoma orbitalis (Shang, 1958; Zhang and Wu, 1987; Wang, 1987; Pan, 1989; Zhi, 1994; Wang et al., 1995; Zhang, 1996; Wang, 1996; Bao et al., 2000; Zhong et al., 2001; Feng et al., 2002; Hou et al., 2003; Zhang et al., 2004a; Zhang et al., 2004b; Yang et al., 2004; Zhang et al., 2005; Guan et al., 2006; Wu et al., 2006; He et al.,

2007; Zhu et al., 2007).

#### **General information**

Sex ratio: 47 cases available in statistics in this study



**Figure 4.** MRI (T2) revealed a cavity-like lesion in tissues.



**Figure 5.** B-type ultrasonography revealed enhanced superficial echo of the mass and rear echo attenuation.

included 25 males and 22 females, with a roughly equal sex ratio. The mean age was  $26.5 \pm 8.8$  years, indicating an earlier onset age.

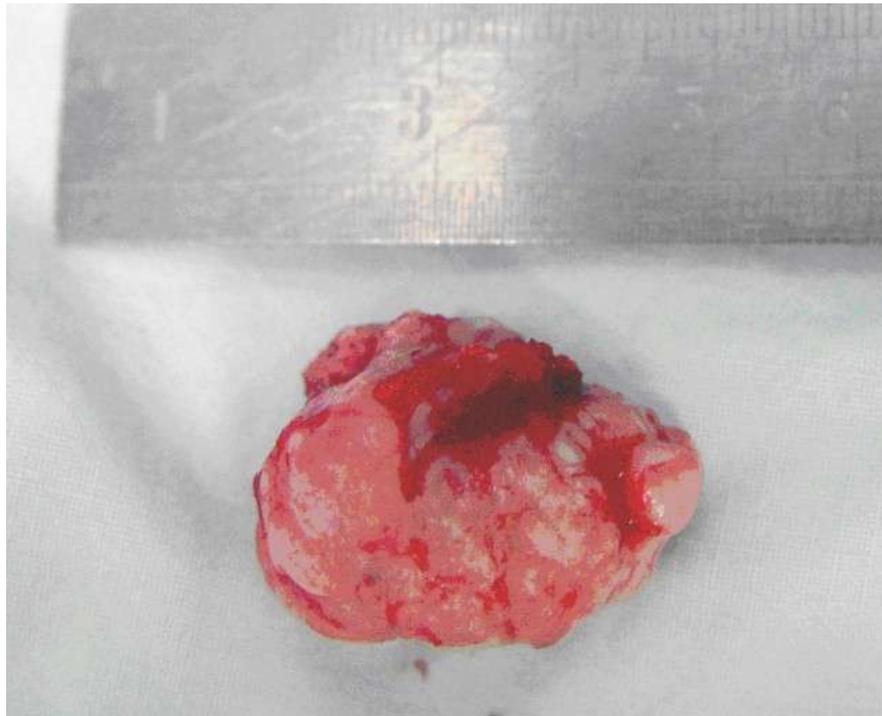
#### Imaging examination

CT examination showed intraorbital tumor presented in

bone density, and the relationship between the tumor and paranasal sinus was also determined. Of 54 cases in reports accounting for 91% of all cases, there were 12 cases in ethmoid sinus, 17 cases in frontal sinus, fronto-ethmoidal sinus in 2 cases, ethmoid sinus and maxillary sinus in one case, sphenoid bone in one case and fronto-sphenoid bone in one case. Besides, sinus cavity was invaded in 6/20 cases with unspecified origins. Due to



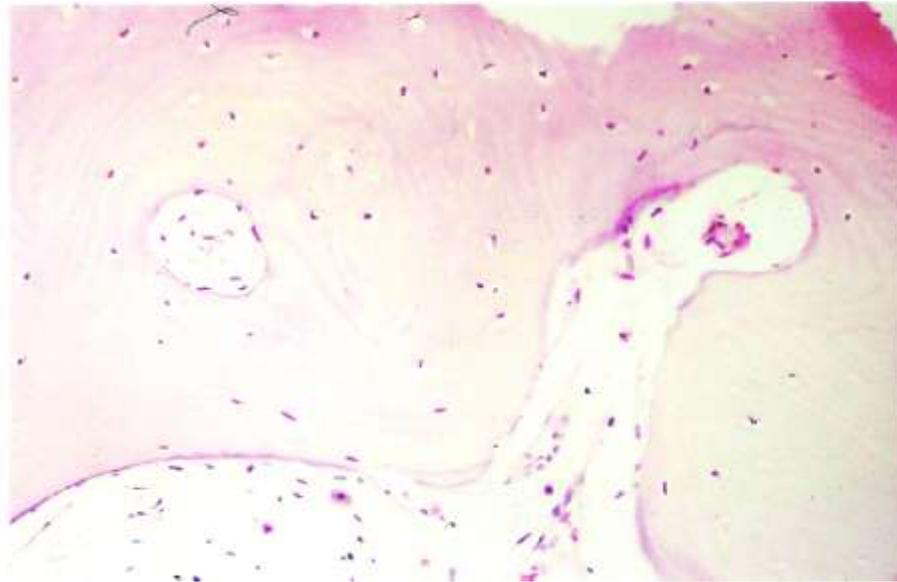
**Figure 6.** The mass was too large to extract through the peri-eyeball space, and thus some superior border of orbital bone was removed.



**Figure 7.** The size of the tumor was 2.4 × 2.8 × 3.2 cm.

poor penetration of B-type ultrasound and color Doppler ultrasound and no blood flow in tumors, they had a little value in the diagnosis of osteoma orbitalis. MRI in 2

cases including one case in this report indicated no tissue signal in T1T2 weighted image in the corresponding sites of tumors, presenting "cavity-like"(Figures 3 and 4), thus



**Figure 8.** Pathology revealed depressed bony surface and bone cells.

MRI in combination with CT had high value in differential diagnosis.

#### ***Literature in Medline***

The first case of osteoma orbitalis was reported by Fridenberg P in 1903 (Fridenberg, 1903). With the title keywords of "Orbita" and "Osteoma", 53 articles were indexed in Medline database, including 25 articles in English and 28 articles in non-English language. Due to non-unified data record, it was hard to obtain full text and perform completely objective statistics, and thus simple overview was performed as follows:

There were a total of 47 cases basically been reported in 53 articles, and the largest sample size was 9 cases reported by El Kohen A in 2005 in French (El Kohen et al., 2005). In tumor-associated clinical conditions, due to difficulty in full text purchase and languages, comprehensive statistical description was not performed. At present, only some available data are listed as follows:

Sex ratio: 11 males and 14 females; females were slightly more than males, similar with the statistical results of literature in Chinese.

Age: 32 years in mean (from 11 years to 65 years)

Paranasal sinus origin or paranasal sinus-related: 44 cases available in statistics included ethmoid sinus/bone-related in 16 cases, frontal sinus-related in 9 cases, ethmoid and frontal lesions in 15 cases, sphenoid in 3 cases and other origin in one case. (Since the above data were obtained from non-full text data, the relative number was not counted in order to prevent from misleading).

#### **Comparison of literature in Chinese index system and Medline**

Notably, there was more detailed and comprehensive description and recordation of body and other eye disorders in foreign literature. Of these 53 articles, it was reported that osteoma orbitalis generated from Gardner's syndrome in two cases (Whitson et al., 1986; McNab, 1998) and Osgood-Schlatter disease in one case (Arseni et al., 1970). Three cases of orbital or paranasal sinus osteoma accompanied with orbital infective inflammation, including one case of fronto-ethmoidal sinus accompanied with orbital subperiosteal abscess (Sahin et al., 2007), one case of orbital osteoma accompanying with dacryocystitis (Benatiya et al., 2006) and sphenoid sinus osteoma accompanied with dacryorrhea and orbital cellulitis (Mansour et al., 1999); one case of osteoma orbitalis associated with gaze-induced amaurosis (Sibony et al., 2004); one case accompanied with foreign substance (Ma'luf et al., 2003). It was recognized that osteoma orbitalis resulted from intraorbital invasion of osteoma of paranasal sinus. Of these 53 articles, a total of 37 articles recorded that osteoma orbitalis resulted from paranasal sinus or intraorbital invasion of osteoma in the paranasal sinus, and systemic diseases, trauma and foreign body were recorded in the remaining 16 articles, significantly different from literature in Chinese (local disease accompanied was reported in only 2/15 cases).

There is a consensus in treatment of osteoma orbitalis in China and foreign countries that surgery is the only effective treatment. However, it is notable that increasing

cases treated by tumor extirpation with nasal endoscopy were reported in foreign literature in recent years (Huang et al., 2001; Naraghi and Kashfi, 2003).

## Conclusion

Osteoma orbitalis is a rare orbital space occupying disease. Zhang et al. (1998) retrospectively analyzed a total of 3406 cases of orbital disease treated in Department of Ophthalmology, second Hospital of Tianjin Medical University from 1976 to 1995, and it was found that there were only 11 cases of osteoma orbitalis, accounting for 0.3%. The majority of osteoma orbitalis was unilateral, with onset age predilection in youth. At present, it is generally recognized that orbital wall is not the primary site of osteoma orbitalis, but it is a benign stromal tumor initially generating from paranasal sinuses due to sclerotization of connective tissues. Inflammatory stimulus and trauma of nasal sinuses might be the etiological factors. However, osteoma orbitalis might originate from embryonic cartilage cells in the junction of frontal and ethmoid bone. There were 3 kinds of pathological types: (1) cortical type (hard type or ivory type): it is hard, small, pedunculated mostly and slowly grows, and it commonly occurs in the frontal sinus; (2) trabecular type (soft type or sponge type): it is porous and fast grows, with large basilar part and volume, and it may generate from fibrous tissues of bones and presents cystic cavity in the center of tumors and hard capsula ossium in the tumor surface, and it commonly occurs in the ethmoid sinus; (3) mixed type: it is hard in the external part while pultaceous in the internal part, and it commonly occurs in the frontal sinus.

The pathological and manifestation changes of osteoma orbitalis are chronic. There are generally no clinical symptoms caused by small tumors while obvious compression symptoms caused by large tumors. If without clinical manifestations, the patients are closely followed up, and the only effective treatment is surgery. Based on the systemic review of literature in Chinese, the clinical manifestations included exophthalmos (76%), periorbital mass (72%) and orbital space-occupying changes, with less impact on visual acuity (47%) and eye movement (40%). Specific changes of osteoma orbitalis could be observed in orbital X-ray and CT, MRI, and contradistinction of CT and MRI might provide more useful diagnostic value. Surgical excision is the main treatment of osteoma orbitalis, but the following features of surgery should be noted.

1) Due to the bony structure, the size and shape of osteoma orbitalis are fixed. The tumor must be fully exposed for complete removal; otherwise tumor compression may lead to eyeball rupture and severe orbital soft tissue injury. Therefore, this problem must be fully considered in preoperative preparation and operation design. Due to the large volume of the tumor in

this patient, some orbital bones in superior margin were removed for complete removal of the tumor.

2) Tumor occurrence and location are highly related with paranasal sinus (91%). Thus, incision and drainage of paranasal sinuses must be fully considered. In theory, simple excision may lead to mucous cysts. Therefore, it will be better that operation design is performed with ENT doctors, and surgical excision could be performed with ENT doctors if necessary. If there are mature techniques and conditions, it is not a bad attempt that nasal endoscopic enucleation for osteoma orbitalis is performed by ENT doctors.

In the course of diagnosis and treatment of osteoma orbitalis, comprehensive and detailed medical history-taking and physical examination must be performed in order to avoid missed diagnosis and misdiagnosis. Tumors that are inside the orbit or next to nose should be identified with osteoma orbitalis. Thus, we must note that osteoma orbitalis generates from paranasal sinuses, and comprehensive medical data and diagnosis should be obtained. Osteoma orbitalis can be show like this: slow onset, with exophthalmos, painless mass next to nose and above eye. If CT showed mass in bone density levels and in MRI showed a vacuole-like structure, osteoma orbitalis should be considered the first diagnosis.

## REFERENCES

- Arseni C, Constantinescu AI, Ghitesco M (1970). Orbital osteoma in Osgood-Schlatter disease. *Presse. Med.*, 78(37): 1630.
- Bao YQ, Wang CY, Yang SB (2000). One case of osteoma orbitalis accompanied with mucocele in ethmoid sinus. *Chin. Ophthal. Res.*, 18(1): 245.
- Benatiya AI, Touiza E, Bhallil S (2006). Orbital osteoma: three case reports. *Bull. Soc. Belge. Ophtalmol.*, (300): 73-79.
- El Kohen A, Lahlou M, Rabeh G (2005). Orbital osteoma: clinical evaluation of nine cases. *Rev. Stomatol. Chir. Maxillofac.*, 106(1): 7-12.
- Feng ZM, Qin JP, Duan QH (2002). One case of osteoma orbitalis. *Chin. J. Med. Imag. Technol.*, 18(3): 207.
- Fridenberg P (1903). Orbital-osteoma of ethmoidal origin. *Trans. Am. Ophthalmol. Soc.*, 10(Pt 1): 83-87.
- Guan XD, Huang CB, Wang Y (2006). One case of large osteoma invading eye sockets, frontal and ethmoid sinuses. *Chin. J. Prac. Ophthalmol.*, 24(11): 1161.
- He W, Luo Q, Tang L (2007). Clinical and pathological analysis of primary orbital bone tumors and tumor-like lesions. *Chin. Ophthal. Res.*, 25(12): 953-955.
- Hou SK, Xiao LH, Wu HY (2003). Four case of osteoma orbitalis. *Chin. J. Ophthalmol.*, 39(1): 54.
- Huang HM, Liu CM, Lin KN, Chen HT (2001). Giant ethmoid osteoma with orbital extension, a nasendoscopic approach using an intranasal drill. *Laryngoscope*, 111(3): 430-432.
- Ma'luf RN, Ghazi NG, Zein WM, Gedeon GA, Hadi UM (2003). Orbital osteoma arising adjacent to a foreign body. *Ophthal. Plast. Reconstr. Surg.*, 19(4): 327-330.
- Mansour AM, Salti H, Uwaydat S, Dakroub R, Bashshour Z (1999). Ethmoid sinus osteoma presenting as epiphora and orbital cellulitis: case report and literature review. *Surv. Ophthalmol.*, 43(5): 413-426.
- McNab AA (1998). Orbital osteoma in Gardner's syndrome. *Aust. N. Z. J. Ophthalmol.*, 26(2): 169-170.
- Naraghi M, Kashfi A (2003). Endonasal endoscopic resection of ethmoido-orbital osteoma compressing the optic nerve. *Am. J. Otolaryngol.*, 24(6): 408-412.

- Pan XF (1989). One case of osteoma orbitalis. J. Fourth. Military. Med. Univ., 9(1): 234.
- Sahin A, Yildirim N, Cingi E, Atasoy MA (2007). Frontoethmoid sinus osteoma as a cause of subperiosteal orbital abscess. Adv. Ther., 24(3): 571-574.
- Shang CX (1958). Eye osteoma: case report. Chin. J. Ophthal., 8: 518.
- Sibony P, Shindo M (2004). Orbital osteoma with gaze-evoked amaurosis. Arch. Ophthalmol., 122(5): 788.
- Wang HH (1987). Large osteoma orbitalis. Chin. J. Prac. Ophthalmol., 5(5): 365.
- Wang JM (1996). One case report of large osteoma orbitali. J. Prac. Radiol., 12(3): 150.
- Wang ZC, Lan BS, An YZ (1995). CT diagnosis of orbital osteoma. Chin. J. Radiol., 29(10): 688-690.
- Whitson WE, Orcutt JC, Walkinshaw MD (1986). Orbital osteoma in Gardner's syndrome. Am. J. Ophthalmol., 101(2): 236-241.
- Wu GH, Wang GG (2006). One cases of primary orbital osteoma. Chin. J. Pract. Ophthalmol., 24(2): 187.
- Yang P, Lee HM, Meng L (2004). Two case of large osteoma orbitalis. J. Injur. Occupat. Dis. Eye. Ophthal. Surg., 26(6): 431.
- Zhang H, Song GX, He YJ (1998). Pathological classification of 3406 cases of orbital diseases. Chin. J. Pract. Ophthalmol., 16(3): 172-174.
- Zhang W, Wu YZ (1987). One case of osteoma orbitalis. J. Jilin. Univ. (Med. Edit), 13(02): 175.
- Zhang WJ, Song GX (2004b). A clinic analysis of 17 cases of orbital osteoma. Chin. J. Prac. Ophthalmol., 22(10): 835-836.
- Zhang XL, Wang JQ, Liu W (2004a). One case of osteoma orbitalis. J. Clin. Ophthalmol., 12(4): 336.
- Zhang XN (1996). Osteoma orbitalis: report of two cases. Chin. J. Ophthalmol., 32(4): 281.
- Zhang ZX, Zhou Y (2005). One cases of large osteoma treated in naso-orbital-orbital approach. J. Clin., Otorhinolaryngol., 19(7): 659.
- Zhi SP (1994). One case of osteoma orbitalis accompanied with orbital cellulitis. Chin. J. Ophthalmol., 30(4): 304.
- Zhong M, Xia FH; Yin RG (2001). One case of large osteoma orbitalis. Chin. J. Ophthalmol., 37(4): 247.
- Zhu SC, Feng GS, Shi DP (2007). CT Diagnosis of Osteogenic Benign Tumor and Tumor Mimics in Orbit. J. Prac., Radiol., 23(12): 1606-1609.