THE USE OF CT IN THE DIAGNOSIS AND DIFFERENTIAL DIAGNOSIS OF ORBITAL MASSES

ORBİTA KİTLELERİNİN TANI VE AYIRICI TANISINDA BTNİN KULLANIMI

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ABSTRACT

This study includes patients with the pre-diagnosis of orbital space-occupying tumor or tumor-like lesions examined on CT in Ankara Training and Research Hospital Radiology Department. Thyroid ophthalmopathy cases were not included in the study.

CT criteria specific to orbital mass lesions and the diagnostic value of CT were investigated through comparison of the CT findings with the histopathological results. Of a total of 40 patients, 22 (55 %) were diagnosed with primary tumors, 6 (12.5 %) with secondary tumors and 12 (32.5 %) with tumor-like lesions.

CT is highly sensitive in the definition of intra-tumoral calcifications, and sclerotic and destructive changes in osseous structures. The location and local invasion of intraorbital space-occupying lesions can be clearly differentiated on CT. Diagnosis of intraocular lesions can be made specifically, correlating CT findings with orbital ultrasound results.
The differential diagnosis of benign and malignant tumoral lesions from inflammatory and vascular lesions can be made using CT.

In some cases where no specific diagnosis can be made, CT directs the therapy by identifying the outlines and morphological characteristics of the lesion. The diagnostic specificity of CT will surely increase in the light of anamnesis, physical examination findings and other radiological modalities.

Keywords: *Orbita, Mass, CT*

ÖZET

Ankara Hastanesi Radyoloji Bölümünde tümör veya tümör benzeri lezyon ön tanısı ile BT tetkiki yapılan ve aynı nedenlerle opere edilmiş olguların BT bulguları incelenerek, tiroid oftalmopati dışında 40 olgu çalışma kapsamına alındı.

Histopatolojik sonuçlarla, BT bulgulan karşılaştırılarak orbital kitlelere spesifik BT kriterleri ve BT'nin tanı değeri araştırıldı. 40 olgunun 22'si primer tümör (%55), 6'sı sekonder tümör (%12.5), 12'si (%32.5) tümör benzeri lezyondu.

Sonuç olarak;

- BT, intratümoral kalsifikasyonların, kemik yapılardaki sklerotik ve destrüktif değişikliklerin saptanmasında çok duyarlıdır.
- BT ile intraorbital yer kaplayan lezyonların lokalizasyonu ve çevre dokulara yayılımı net olarak belirlenebilmektedir.
- İntraoküler lezyonlar, USG ile korele edildiğinde spesifik tanılara ulaşılabilmektedir.
- Orbital BT ile benign ve malign tümoral lezyonlar ile inflamatuar ve vasküler lezyonların ayıcı tanısı yapılabilmekte, spesifik tanıya gidilemeyen olgularda, BT lezyonun sınırlarını ve morfolojik özelliklerini belirleyerek tedaviyi yönlendirmektedir.

BT ile spesifik tanı konabilme oranı; BT bulgularının anamnez, fizik muayene ve diğer radyolojik görüntüleme yöntemlerinin sonuçlarıyla beraber değerlendirildiği ölçüde artış gösterecektir.

Anahtar Kelimeler: *Orbita, Kitle, BT*

INTRODUCTION

Ultrasonography (US) and Computed Tomography (CT) have been used as important diagnostic methods in the evaluation of orbital masses. With the use of these methods, it has become easier to define an orbital mass with all its properties, even in cases where the clinical and laboratory findings are not sufficient [1, 2].
The configuration, localization, and density properties of masses, intracranial/facial invasions, and the condition of the orbital bones can be easily evaluated using CT, thereby increasing the likelihood of a correct diagnosis [3].
The aim of this study was to define specific CT characteristics of orbital masses, and to evaluate the success of CT in the differentiation of masses through a comparison of the CT images and pathological/ clinical diagnosis of the included patients.

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MATERIALS AND METHODS

Approval for this study, which was conducted in Ankara Education and Research Hospital, was granted by the hospital Ethics Committee. A retrospective examination was made of the records of patients who were operated on by the Ophthalmology or Brain Surgery Departments for a diagnosis of orbital tumor in a 2-year period. The patients included had a suitable CT scan available. Cases of thyroid ophthalmopathy were excluded. A total of 40 patients were included in the study.

CT examinations were performed with a Hitachi W 950 SR machine, with slice thickness of 2 mm for axial and 3 mm for coronal slices (mAs 140 Ah, kV 120 V). The orbita was scanned from the superior wall of the maxillary sinus to the floor of the frontal sinus, parallell to the infraorbital line, at an angle of 10˚ to the orbitomeatal line. Non-ionic contrast material at a dose of 0.4ml/kg was injected intravenously before the scan. Soft tissue and bone filters were used.
The localization of the lesion was described as intraocular, intraconal with/without optic nerve involvement, extraconal-intraorbital, extraconal-extraorbital, lacrimal gland.

CT findings were recorded as localization, density (containing oil, cystic, isodense to muscle), presence of contrast enhancement, contour characteristics, presence of calcifications, intracranial/facial invasion and associated bone changes. US findings and plain radiograph findings were also noted, if present.
The parameters were correlated with histopathological results in order to define the CT characteristics of each lesion.

Statistical analysis

All study information was recorded on patient data sheets, then entered into an Excel (2007, Microsoft Corp., Redmond, WA) spreadsheet for analysis. All data entries were double-checked by one of the investigators. Data were analyzed using Statistical Package for the Social Sciences (SPSS) for Windows 20 software (IBM SPSS Inc., Chicago, IL, USA).

Normal distribution of the data was evaluated with the Kolmogorov-Smirnov test. Numerical variables with normal distribution were stated as mean ± standard deviation (SD) values.

Variables not showing normal distribution were stated as median values (interquartile range). For comparison of the numerical variables between the two groups, the Student's T test and Mann-Whitney U test were used. The ANOVA and Kruskall Wallis H tests were applied for comparison between three or more groups. To evaluate the categorical variables, the Chi-Square and Fisher's Exact Chi- Square tests were used. Pearson and Spearman correlation analyses were applied to evaluate the relationships between numerical variables.

A value of $p<0.05$ was considered statistically significant.

RESULTS

Evaluation was made of a total of 40 patients including 10 pediatric patients (0-10 years, 37.5%), and 25 adults (62.5%). The patients comprised 18 females (45%) and 22 males (55%) with a mean age of 31 years (0-65 years). The distributions of the pathological diagnoses according to age groups are shown in **Tables 1 and 2.**

Table 1: Age groups

Age groups (years)	Male	Female	Total	Percentage
0-10	9	6	15	37.5
11-20	2	2	4	10
21-30	3	1	3	7.5
31-40	4	2	6	15
41-50	2	1	3	7.5
51-60	2	2	4	10
61-70	1	4	5	12.5

Table 2: Pathological diagnosis according to age subgroups

Age groups (years)	Number (percentage)	Diagnosis
0-10	15 (7.5%)	Optic glioma, neuroblastoma, leukemia metastasis, eosinophilic granuloma, rhabdomyosarcoma, neurofibromatosis, lymphangioma, dermoid and epidermoid cysts, fibrous histiocytoma, retinoblastoma
11-20	4 (10%)	dermoid cyst, fibrous dysplasia, orbital cellulitis
21-30	3 (7.5%)	Menengioma, epidermoid cyst, sphenoid wing meningioma
31-40	6 (15%)	Neurofibroma, cavernous hemangioma, metastasis to bulbus oculi, optic glioma, ciliary melanoma
41-50	3 (7.5%)	Metastasis,subperiostal hematoma
51-60	4 (10%)	Pseudotumor, angiofibroma, paranasal sinus neoplasm, retrobulbar abscess
61-70	5 (12.5%)	Lymphoma, mucocele, uveal melanoma

The reasons for presentation at hospital were exophthalmia (25%), periorbital inflammation (20%), diplopia (5%), loss of vision (15%), and white pupil – strabismus (7.5%).
Pathological findings observed on plain radiographs were of cranial meningioma extending into the orbita, neuroblastoma metastasis, eosinophilic granuloma,

neurofibromatosis, and subperiosteal hematoma **(Figure 1, 2 and 3).**

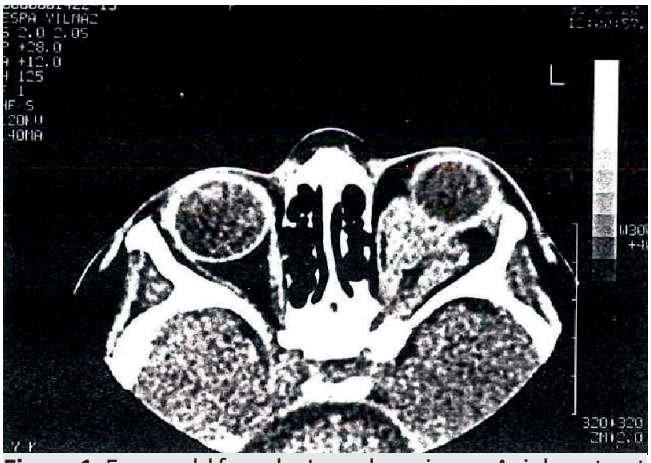

Figure 1: 5 year old female. Lymphangioma. Axial contrast enhanced CT image showing infiltrative, non enhancing, lobulated mass at retrobulber area on the left.

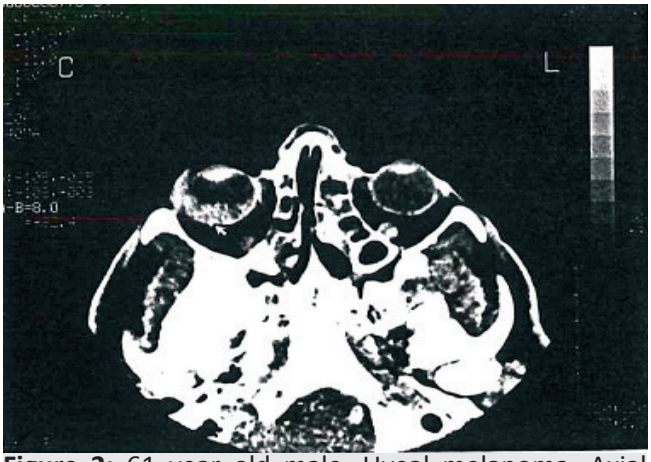

Figure 2: 61 year old male. Uveal melanoma. Axial contrast enhanced CT image showing homogenously enhancing cresentric mass on the right.

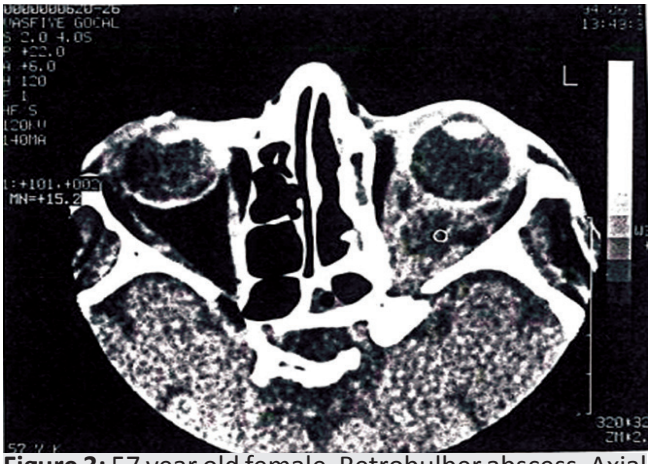

Figure 3: 57 year old female. Retrobulber abscess. Axial contrast enhanced CT image showing peripherally enhanced, cystic intraconal mass on the left.

The lesions were bilateral in 39 (99%) cases, and unilateral in 1 case (1%-retinoblastoma). Details of the localizations of the lesions are shown in **Table 3.**

Table 3: Pathological diagnosis according to localisation

Localisation	Number (percentage)	Diagnosis
Intraocular	8 (20%)	Metastasis to bulbus oculi, retinolastoma, uveal and ciliary melanoma
Intraconal	7 (17.5%)	Cavernous hemangioma, optic glioma, meningioma, pseudotumour, retrobulbar abscess
Extraconal	2 (30%)	Dermoid cyst, rhabdomyosarcoma, orbital cellulitis, neurofibroma, epidermoid cyst, subperiostal hematoma, angiofibroma
Intra + Extraconal	4 (10%)	Lymphoma, neuroblastoma, lymphangioma
Extraconal+ Extraorbital	9 (22.5%)	Fibrous dysplasia, fibrous histiocytoma, paranasal sinus neoplasm, metastasis, mucocele, eosinophilic granuloma, neurofibromatosis, sphenoid wing meningioma

The density was equal to fat in 2 cases of dermoid cyst, and equal to water in 3 cases of epidermoid cyst. Dense liquid/cystic densities were found in cases of orbital cellulitis, retrobulbar abscess, and mucocele. In a fibrous dysplasia case, bone density was determined and in all the other cases the density was similar to that of muscle.

Diffuse contrast enhancement was seen in 5 cases (12.5%): cavernous hemangioma [2], meningioma [2], angiofibroma [1]. Peripheral contrast enhancement was present in cases of dermoid and epidermoid cyst, mucocele, orbital cellulitis, and retrobulbar abscess. Mild contrast enhancement was present in cases of rhabdomyosarcoma, lymphoma, metastasis (neuroblastoma, leukemia, leiomyosarcoma), fibrous histiocytoma, and neurofibroma. There was no contrast enhancement in 2 cases of subperiosteal hematoma.

Smooth contours were observed in cases of cavernous hemangioma, dermoid-epidermoid cyst, angiofibroma, and mucocele, whereas irregular-indistinct contours were seen in cases of lymphangioma, lymphoma, pseudotumor, metastasis, and inflammatory/infectious lesions.

Calcifications were determined to be present in 6/40 (15%) lesions: meningioma (2- amorphous diffuse), optic glioma (1-puctate localized), retinoblastoma (3-noduler localized) **(Figure 4).**

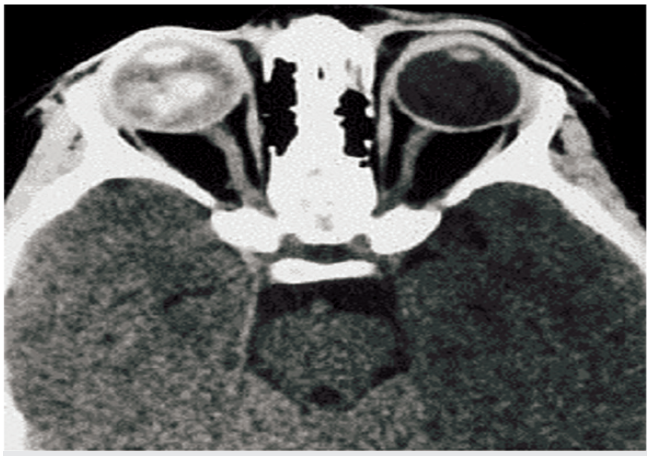

Figure 4: Retinoblastoma. 5 year old boy. Axial CT image without contrast medium injection. There is a calcified mass filling most of the vitreus.

Associated bone lesions (destruction, erosion, sclerosis, expansion, enlargement of optic canal) were present in 16/40 (40%) lesions **(Table 4)**

Table 4: Associated bone changes

Bone changes	Number of cases	Lesions
Destruction	7	Lymphoma, metastasis, rhabdomyosarcoma, eosinophilic granuloma, paranasal sinus neoplasm,
Erosion	2	Mucocele, epidermoid cyst
Sclerosis	2	Meningioma, fibrous dysplasia
Expansion	3	Fibrous histiocytoma, dermoid- epidermoid cyst
Enlargement in optic canal	2	Optic glioma, meningioma

Intracranial extension was determined in 5/40 (12.5%) lesions: lymphoma, neuroblastoma metastasis, optic nerve glioma, paranasal sinus neoplasm, and sphenoid wing meningioma. Intraorbital extension from an intracranial mass was observed in one case of middle cranial fossa meningioma.

DISCUSSION

Orbital masses can mimic each other in respect of clinical and laboratory findings. Exophthalmia, periorbital edema, loss of vision, pain with eye movements or eye movement defects, and palpable mass are some common manifestations. As previously stated, the history and physical examination of the patient are often insufficient for the diagnosis of orbital masses, so imaging methods are frequently used [1, 2, 4].

Plain radiographs can show some changes such as increased soft tissue thickness, orbital expansion, bone destruction, sclerosis, changes in the optic foramen, and intra-tumoral calcifications. US is limited in the identification of bone changes and is user-dependent. Angiographic methods can be used to diagnose vascular lesions [5-7].

CT is utilized in the diagnosis of orbital masses, and assessment of orbital bone changes. It is a useful method with high imaging resolution and multiplanar imaging capacity [7].

In the current study, it was aimed to define the CT findings for discrimination of orbital masses using the 40 cases included.

The incidence of orbital masses differs amongst studies. Generally, in a pediatric population, capillary hemangioma, lymphangioma, rhabdomyosarcoma, dermoid-epidermoid cysts, optic nerve glioma, leukemia, neurofibroma, metastatic neuroblastoma, and retinoblastoma are commonly seen. Whereas, in adults, orbital cellulitis-abscess, cavernous hemangioma, orbital lymphoma, pseudotumor, dermoid-epidermoid cysts meningioma, metastasis and lacrimal gland tumors are commonly diagnosed [3, 8-11].

Optic nerve glioma and meningioma are the primary lesions located in the intraconal area.

Neurinoma is another benign peripheral nerve tumor which encircles the optic nerve and can be seen intraconally. In the current study, optic nerve involvement was seen in 3 cases, and it presented as enlargement and contrast enhancement of the optic nerve. There were 2 optic glioma cases. One was pediatric, and in this case the tumor filled the suprasellar cistern and involved the optic chiasm. The adult case showed a rare finding of intratumoral calcification.

CT is the primary choice to evaluate bone changes, and the presence of calcifications, but it is relatively weak in showing intracranial/intracanalicular extensions, for which magnetic resonance imaging (MRI) should be applied. MRI was used to support the CT diagnosis in the pilocytic astrocytoma case of this study. Hemangioblastoma, hemangiopericytoma and schwannoma must be kept in mind in the differential diagnosis [12-15].

Primary orbital meningiomas constitute 5% of primary orbital tumors. Intracranial meningioma extending into the orbita is rarer. The meningioma case in this study contained coarse and diffuse intraorbital calcifications. Sclerosis in the optic foramen and a concurrent middle cranial fossa mass were other clues for the diagnosis.

Although the presence of a central radiolucent line has been defined for meningiomas, it may also be seen in optic neuritis and pseudotumor. This finding was not detected in this case [16, 17].

Three cases with intraconal compartment localization but which did not involve the optic nerve were cavernous hemangioma and lymphangioma. Cavernous hemangiomas are seen as intraconal, circumscribed masses with avid enhancement, and rarely contain calcifications [4, 18, 19].The CT findings of the current study were consistent with the literature. Meningioma, hemangiopericytoma, and schwannoma might mimic these masses, in which case MRI can be useful in differentiation [19]. Lymphangiomas tend to show progression, and present as diffuse, infiltrative masses without contrast enhancement. In contrast, hemangiomas are more circumscribed in shape [20]. The findings of the lymphangioma case of the current study were similar to findings reported in literature.

The retrobulbar-intraconal lesions found in the current study were pseudotumors. They generally present with heterogeneity in retrobulbar fat with obscured margins. Sometimes pseudotumor cannot be diagnosed confidently with imaging, and in these cases histopathological sampling might help. Pseudotumor cases can mimic thyroid ophthalmopathy and lymphoma [21, 22].Orbital myositis must be differentiated from dacryoadenitis, periscleritis, perineuritis, and diffuse cellulitis cases [23].

Extraconal-intraorbital compartment lesions tended to be dermoid-epidermoid cysts, rhabdomyosarcoma, and neurofibroma in the current study. Dermoid-epidermoid cysts are generally found in the frontozygomatic fissure line. They present as isodense to fat, generally. They have regular contours, and cause cortical thinning to the adjacent bone [24, 25].

Rhabdomyosarcoma is the most common sarcoma type for pediatric patients with a mean age of 8 years. Embryonal rhabdomyosarcoma is the most common subtype. They often cause extra orbital extension and bone destruction. Therefore, differential diagnosis must be performed between neuroblastoma metastasis and Ewing's sarcoma. Moreover, as they present as rapidly growing masses it is difficult to separate rhabdomyosarcomas from cellulitis [26]. In the rhabdomyosarcoma cases of the current study, solid masses were detected that could not be easily differentiated from periorbital muscles, and the mass caused bone destruction. In another case included in the study, it presented as a rapidly growing, irregularly shaped, heterogeneously enhancing mass encircling the orbit from the superomedial side.

Neurofibroma and schwannomas originate from Schwann cells of peripheral neurons. They can be seen isolated or associated with neurofibromatosis.Isolated forms might mimic hemangioma, meningioma and hemangiopericytoma with similar morphological characteristics and enhancement patterns on CT images. The cases that present as solitary masses on CT can be multifocal [27].In the current case, the typical signs of sphenoid wing dysplasia, thickening in periorbital soft tissues- palpebral, and herniation of the temporal lobe were all present. The diagnosis was confirmed pathologically.

Orbital metastasis constitutes 3-9% of orbital masses. In adults, orbital involvement is 1/7, and in the pediatric population, orbital involvement is more common. Neuroblastoma and Ewing's sarcoma are the most common metastasis types in children. Breast, lung and gastrointestinal malignancies, in increasing order, are the most common metastasis in adults.

It is not very uncommon to detect orbital metastasis before the tumor itself. In the metastasis cases of the current study, bone destruction (mainly at sphenoid), and soft tissue mass were determined in primary leiomyosarcoma and pancreatic adenocarcinoma cases. In neuroblastoma metastasis, irregular bone destruction, and intracranial/intraorbital extension is present, and the mass is hyperdense. The hyperdensity is important for differentiation between rhabdomyosarcoma and neuroblastoma metastasis. In addition, neuroblastoma metastasis does not tend to involve preseptal soft tissues. Acute myelomonocytic leukemia has been defined as creating metastasis to the orbit, as a retrobulbar, infiltrative, enhancing mass. It is important to differentiate metastasis and primary orbital lymphoma [28, 29].

Extraconal-extraorbital lesions were determined as fibrous dysplasia, mucocele, sphenoid wing meningioma, and subperiosteal hematoma, consistent with the literature [1, 2].

Orbital lymphoid lesions have been found at rates between 11-20% in pathology series. Isolated malign lymphoid orbital neoplasia is seen rarely. Differential diagnosis includes malignant melanoma and multiple myeloma. In some cases, biopsies can be the only means of discrimination [2].A component of the systemic disease is more commonly seen than primary cases. In the current case, the orbital lymphoma presented as a palpable mass with a long standing proptosis. The mass was mildly enhanced and bone destruction, and intracranial extension were also present.

Eosinophilic granuloma is a component of the Langerhans cell histiocytosis spectrum. They can be seen as solitary or multiple masses. In orbital bones, the frontal and lateral orbital wall are the most common areas to be affected. Characteristically, they are seen as sharply delineated lytic lesions and concurring soft tissue mass [30]. The current cases were identical with the literature.

Orbital fibrous histiocytoma is a mesenchymal origin tumor, which can be benign or malignant. It can be seen between the ages of 4-84 years and is generally located superomedially. It presents with loss of vision, proptosis and a palpable mass. On CT, they are expansile, enhancing, well-shaped lesions [31].The single case of fibrous histiocytoma in the current study was consistent with the literature.

Paranasal sinus malignancies are a common pathology extending into the orbita. The most common type is maxillary sinus neoplasia with an incidence of 40-65% intraorbital extension. Demonstrating the orbital extension is important for the treatment plan [32].In the current case, the neoplasia filled the nasal cavity, ethmoidal cells, and maxillary sinus. It extended into the orbita with destruction of the medial and inferior walls of the orbita.

The most common primary intraocular tumor in the pediatric population is retinoblastoma. It is generally seen under the age of 5 years. The presence of calcifications is very common and when calcifications are absent, it is difficult to differentiate from primary persistent hyperplastic vitreous, Coat's disease, retrolental fibroplasia, retinal astrocytoma, chronicle retinal detachment, and Toxocara infestation. In this case MRI can be problem solving [33].

The current study cases presented with calcifications and mild contrast enhancement extending into the uvea

Malignant melanoma is the most common primary tumor of the orbita. Diagnosis of ciliary melanomas in particular is CT and MRI dependent. MRI is superior to CT in differentiating hemorrhagic and serous choroidal detachment [33].

CONCLUCION

Intra-tumoral calcifications can be seen in hemangioma, lacrimal gland tumors, optic glioma and meningiomas, but they mainly indicate meningiomas.

Fat densities are characteristic for dermoid cysts. Avid contrast enhancement is seen in hemangiomas, and neuroblastoma metastasis. Peripheral contrast enhancement is seen in mucocele and abscess cases. The absence of enhancement points to mucocele, hematoma and dermoid cyst.

Cortical thinning and fossa formation indicate dermoid cyst. Hyperostosis is generally seen with meningiomas.

Enlargement of the optic canal indicates optic glioma, and if seen together with hyperostosis, then meningioma is the correct diagnosis. If bone expansion and erosion are related with paranasal sinuses, mucocele should be considered. Irregular and diffuse destruction generally indicates malignancies, such as metastasis, lymphoma, and rhabdomyosarcoma.

Prominent expansion and sclerosis without destruction generally indicate fibrous dysplasia.

CT is a helpful method in the diagnosis and differentiation of orbital lesions. In addition, it can guide clinicians in determining and planning the proper treatment.

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