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MODERN DIAGNOSIS OF ORBITAL TUMORS

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The advent of echography (especially quantitative A-scan) and of computerized axial tomography has revolutionized our approach to orbital tumors. We are now in a position to diagnose most orbital tumors not only as to their site and size, but also to their histologic characteristics. This has also changed our management of these lesions. While in previous years an exploratory biopsy was frequently necessary to establish the diagnosis of a tumor or to determine its histologic character, this is at present hardly necessary any more. The reliability of our diagnostic tests is so high that we can make the clinicial diagnosis with a great deal of accuracy. The only exception is the group of lymphoid tumors in which the differentiation between a benign hyperplasia and a malignant lymphoma is clinically still difficult. In most of these instances a biopsy will be neccessary even though the pathologic picture is frequently inscrutable.

1. *GENERAL EXAMINATION*

It is understood that any patient with an exophthalmus needs a general physical examination and a good history. It is of importance whether the exophthalmus came on suddenly or whether it was present for many years. We have to know whether it is associated with orbital pain or not. Wet also should know whether the patient suffers from any systemic diseases, especially from a thyroid disorder.

The examination comprises not only a complete ocular examination, but also an accurate evaluation of the exophthalmus. We have to determine whether the eyes are pushed straight forward or whether there is also a lateral or vertical deviation. We have to find out whether and to what

extent the ocular motility is impaired. Obviously, the fundus examination has to consider pressure on the optic nerve or on the blood vessels and this has to be tested also by determining visual acuity and visual fields. Of special interest is an accurate and vigorous palpation of the orbit in which we push the little finger as deep into the tissues as possible to find out whether any mass can be palpated and whether its consistency can be evaluated.

2. *X-RAY DIAGNOSIS*

Obtaining adequate x-ray pictures will be the next step in the diagnostic procedure. Plain x-rays of the orbit, optic canals and the skull are necessary to evaluate bony changes or certain pathologic changes of the adjacent organs. While these x-rays are the basis for further examinations, the results are rarely positive. In less than 10% of patients with exophthalmus do we find anything of significance on plain x-rays.

If, however, there is a suggestion or suspicion of a bony lesion, tomographic pictures should be taken. This will reveal such changes in much more detail and will allow a precise localization.

Invasive radiologic tests are usually unnecessary and obsolete for the diagnosis of orbital pathologic processes. In the past we used to perform orbitographies (injection of air or radiopaque material into the orbit) or phlebographies (injection of radiopaque material into the angular or facial veins) in order to visualize the intraocular tumor. Indeed these methods will frequently outline the size and the site of the lesion. Orbitography, however, has a certain amount of morbidity as the material can be injected subdurally and has therefore been abandoned. Phlebography many also reveal a filling defect, but not only is this method painful and cumbersome, we learn nothing about the etiology of such a defect. It could be caused by pressure of a mass (the nature of which remains undetermined) on the veins or by an intrinsic process in the veins (such as phlebitis) which would not allow the passage of radiopaque material.

Arteriography is only rarely indicated as a diagnostic tool for orbital lesions. We occasionally do an arteriogram when we assume that a vascular anomaly of unusual nature or one that could extend into the intracranial cavity is the main pathologic orbital process. Is general an arteriography will only be necessary in 1-2% of all orbital lesions.

3. *ECHOGRAPHY AND CAT-SCAN*

These two methods have revolutionized our diagnostic armamentarium.

Echography, i. e. the examination of echoes from ultrasound waves of a wavelength of 6-10 MHz, allows us to examine soft tissues in a fashion that x-rays allow us to examine bony structures. The importance of this new method cannot be overestimated. This is especially suitable for ophthalmology as no bony structures interfere and all ocular and orbital structures are easily accessible to this method of examination. We distinguish the A-scan which shows us amplitudes (or vertical deviations) against time (or distance). This method allows more versatile examinations and gives quantitative results. It is, however, more complicated to read and needs a certain amount of training and practice. The B method, on the other hand, gives us cross sections through the area to be examined and allows therefore a more visual and better topographical evaluation of the lesions.

Computerized tomography has added another valuable dimension to our diagnostic armamentarium. This method is still in the stage of refinement and thinner and better cross sections are becoming available. The method has the advantage that it is usually done by experienced radiologists and needs neither special technicians nor an experienced ophthalmologist for interpretation. It does, however, at the present time not give all the quantitative information we get from the A-scan.

4. *GRAVES'DISEASE*

While the diagnosis can usually be made quite easily just on the basis of the clinical picture, we do have patients in whom the condition is either atypical or unilateral. In these instances we have to be sure that there is no space-occupying lesion in the affected orbit. Here the echographic examination is most valuable. It can be done quickly without any discomfort or deleterious effect and it may once and for all establish the diagnosis of a Graves'disease.

It is also of great importance to realize that even in patients who clinically look like an absolutely unilateral exophthalmus, echographic examination of the apparently normal other orbit will usually reveal thickened extraocular muscles, the hallmark of Graves'disease.

The thickened extraocular muscles can also be seen on the CT-scan. We have to be cautious, however, not to confuse these muscles with a tume-

faction. This is especially prone to occur at the apex where the muscles converge. Additional sections, somewhat higher or lower, will usually reveal the true nature of the mass.

Echographic examination will reveal, first of all, the thickening of the orbital tissue, i. e. an edema of the adipose tissue. This will enlarge and widen the echo from the orbit. Again even the apparently uninvolved eye may show such a widening of the orbital echo, though of a leeser degree.

Nearly pathognomonic is a diffuse thickening of the extraocular muscles which is sometimes accompanied by a thickening of the sheaths of the optic nerve. These findings can be seen on A-scan and B-scan.

The histologic features of this disease explain the echographic and CT findings. We usually find edema in the adipose tissue and a diffuse edematous lymphocytic infiltration of striated muscle.

Occasionally we find a thickened muscle that shows such a massive tumefaction that exploration and surgical excision may be necessary.

5. *HEMANGIOMA*

The hemangioma occurring in adults is usually a cavernous one. The echographic pattern is highly characteristic and this may in some cases make an operation unnecessary.

Clinically characteristic are the slow onset and the location within the muscle cone. This may produce not only exophthalmus but pressure on the globe with the development of choroidal folds and induced hyperopia.

On the CT-scan the lesion is well visible and appears as a sharply outlined, spherical or ovoid structure.

The true nature of the lesion can be appreciated on echography. Characteristic is the location within the muscle cone, the high acoustic reflectivity, the fact that it is not compressible and the high sound attenuation.

Histologic examination explains the echographic pattern. We find numerous connective tissue septa dividing endothelial-lined spaces. This causes strong reflections because we find large interfaces separating areas of different acoustic densities.

The hemangiomas in children and infants ase somewhat different in behavior and clinical course. These hemangiomas are usually capillary or

angioplastic. They are richly vascularized and frequently have a positive Doppler phenomenon which means that they have an active, strong blood flow. They often lie outside the muscle cone.

Now that the diagnosis of an orbital hemangioma can be made with a high degree of certainty, surgical excision should only be performed when really indicated and not for diagnostic purposes only. An adult patient with a mild exophthalmus and no other ocular complaints may not need any surgical procedure, only a follow-up examination at yearly intervals. We suggest surgical removal of such a benign hamartoma only when there is interference with ocular function or it represents a cosmetic blemish.

6. *NEUROGENIC TUMORS*

a. *Meningioma*

These lesions are frequently diagnosed on plain x-rays because of the secondary hyperostosis around the sphenoid ridge. However, if the tumor is only intraorbital, such osseous reactions will not occur. In these instances the CAT-scan will show a diffuse thickening of the optic nerve.

On echography the swollen optic nerve will be seen surrounded by thin sheaths.

Histologic examination reveals a neoplastic proliferation of arachnoidal endothelial cells with a strong tendency to invade bony tissues.

b. *Optic nerve glioma*

This lesion is characterized not only by a slowly progressing exophthalmus, but also by poor vision, optic atrophy and frequently by an enlargement of the optic canal.

The CT-scan will again show a massive widening of the optic nerve.

The echo scan will reveal a diffuse thickening of the optic nerve with a conspicuous enlargement of the meninges. The optic nerve itself has low to medium optic reflectivity. The topography of the lesion can be well demonstrated on B-scan.

In general, an excision of the tumor is not warranted. In young patients these lesions are multiple hamartomas which should not lead to any dif-

ficulty. However, occasionally a biopsy may be necessary as a differentiation from a meningioma may be difficult or impossible. Meningioma in a young patient, on the other hand, is a most aggressive and potentially dangerous condition. The biopsy can be performed through a lateral orbitotomy.

Excision of the tumor will only be necessary if the eye is blind and the exophthalmus becomes a cosmetic blemish. The eye itself can usually be saved.

The eye will, of course, be blind and a severe pigmentary atrophy of the retina will follow.

Histologic examination shows a diffuse proliferation of astrocytes replacing the normal axon cylinders.

7. *RHABDOMYOSARCOMA*

This is the most frequent primary malignant orbital tumor in children. Clinically, it is characterized by sudden onset, rapid progression and the occurrence of hemorrhages.

On CT-scan the entire orbit will be shown to be full of a tumor mass.

On echography the tumor consists of a rather homogenous infiltration and therefore shows low acoustic reflectivity. Hemorrhages may be interspersed.

A biopsy is usually necessary to prove the nature of the tumor. It may be difficult to find viable tumor cells in large areas of necrosis and hemorrhage. The neoplastic cells resemble striated muscle cells.

8. *LYMPHOMA, LYMPHOID HYPERPLASIA AND INFLAMMATORY PSEUDOTUMOR*

It is frequently difficult to distinguish these three entities on clinical examinations alone. All three of them are characterized by a diffuse, small cell infiltration into the orbital tissues.

Echographic examination reveals a lesion of low acoustic reflectivity. This is due to the fact that these infiltrates consist of a rather uniform cell mass without any interfaces of septa which could produce an echo. The lesion may have a sharp outline or extend diffusely in adjacent tissues.

Neither the echographic examination nor the CAT-scan can distinguish cell types. This is beyond the resolution power of these examination methods. We therefore can make no conclusions as to the nature of the small cell infiltrate and a biopsy will often be necessary.

9. *METASTATIC CARCINOMA*

Metastatic lesions to the orbit often come from breast cancers in women or from gastrointestinal or lung cancers in men.

The echographic pattern is quite typical and shows the characteristic V-pattern. This is due to the fact that in the center of the lesion we find a rather uniform infiltration of the orbital tissues by carcinoma cells. In the periphery of the lesion, on the other hand, we find high acoustic reflectivity because in these areas there is an admixture of neoplastic infiltrates with pre-existing orbital tissues, such as connective or adipose tissue.

10. *MUCOCELE*

Here the clinical picture may be quite characteristic. Most of the time the orbit is invaded by a mucocele from the frontal sinus, but it may also come from the ethmoidal cells or the maxillary simus. The picture may occasionally be misleading as for instance in a patient we saw recently with bilateral frontal sinus mucocele that occurred for enough behind the septum so that the lesion couldt not be seen nor palpated and the clinical picture resembled that of a Graves' disease orbitopathy.

While the x-ray examination will usually reveal the correct diagnosis, the echographic pattern is also quite characteristic. We usually find a sharply demarcated defect in the bone. The mucocele itself appears as a cyst-like structure which is barely compressible and has rather homogenous contents and therefore a low acoustic reflectivity.

CONCLUSIONS

If we compare the reliability of echographic diagnosis, we find that the standardized A-scan has an accuracy which exceeds 90%. This compares to an accuracy of 80% with immersion B-scan or of 60% with nonstandardized A-scan examination.

In the hands of Dr. Ossoinig, who is in charge of our echographic laboratories, we are able to detect an orbital tumor in 99% of the cases provided that its size exceeds 1 mm, in the anterior orbit, 2 mm in the posterior orbit and 3 mm at the orbital apex.

We are able to come up with the correct differential diagnosis or tissue diagnosis of an orbital lesion in 85% of the cases by using the standardized A-scan method. In order to obtain this, the orbital lesion has to exceed 4 mm in diameter in the anterior orbit, 8 mm in the posterior orbit, and 10 mm at the orbital apex.

In a recent comparison of 27 consecutive, unselected cases of orbital tumor, we came up with the correct diagnosis by A-scan echography in all of these lesions and by CT-scan in 23 patients.

In summary, we have to say that both methods have advantages and disadvantages. The advantages of the CT-scan are that it gives a better topographic image and allows us to examine retrobulbar structures and the orbital walls. It also allows us to evaluate peri-and retroorbital regions, such as the periorbital sinuses or the intracranial cavity. It is an easier examination technique and it is more convenient to evaluate the results. In addition, the technology of this method is constantly advancing and more advantages can be expected.

On the other hand, the advantages of echography over the CT-scan is, first of all, that echography is an absolutely harmless method that can be repeated again and again and can be used on small children and pregnant women with impunity. This is in contrast to the amount of x-rays used in the CT-scan. In addition, echography is at the present more sensitive and reliable in detecting small tumors and establishing tissue diagnosis. Echography also has the advantage that the equipment is mobile and therefore can be brought into the operating room. Echographic examinations are done by ophthalmologists or by and ophthalmic technician. We therefore have close coordination between the surgeon and the examiner. In addition, the cooperation needed from the patient is much less than that needed for CT-scan. Finally, echographic equipment is much less expensive than a CT-scan.

We therefore have two examination methods available which we should use judiciously and complementarily and follow up with a biopsy if necessary. However, in most of these cases we are now able to establish the presence of a tumor and its nature by using noninvasive examination methods preoperatively.