

Le centre antirabique de l'Institut Pasteur de l'Iran (Téhéran; 69 Avenue Pasteur) est jour et nuit à la disposition des blessés par les animaux suspects, et les blessés sont priés de se présenter immédiatement après la morsure à cet Institut ou, dans les provinces, aux services sanitaires .

RESUME

La névrose rabiforme ou " fausse rage " n'est pas une simulation mais bien le résultat d'une psychose vraie, très grave dans ses conséquences (danger de suicide) .

Plusieurs observations de cette névrose sont soumises ici à l'attention des confrères et spécialement des psychiatres qui possèdent maintenant les moyens de traiter ce type de névrose.

SUMMARY

The Rabiform Neurosis or " False Rabies " is not a simulation, but it is the result of true psychosis, The consequence of which is very severe (danger of suicide).

Here, several observations of this neurosis are brought to the attention of the Colleagues, specially the Psychiatres who, now, have the facilities to cure this type of Neurosis.

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ANEURYSMAL BONE CYST Report of four cases

A. Modjtabai* S. Sarkisian** H. Saleh***

This is a cystic lesion situated under the periosteum usually in the metaphysis of the long bones, and has radiologically a " Blown - out " appearance. Histologically it consists of vascular fibrous tissue. The vessels have no muscle in their walls and no aneurysmal changes.

In 1942 Jaffe and Liechtenstein (5) gave a comprehensive description of the lesion and coined this name on radiological and histological ground.

Before Jaffe described this lesion, many cases were diagnosed and treated as malignant or giant cell tumors of the bone (4).

Since Jaffe and Liechtenstein gave a full account of the lesion there has been many publications on this subject. (Bhaskar et al. 1950 (2) . MacCarty et al. 1961 (7) . Donaldson 1963. Subramanian and Mathias(8) . Ebling and Wagner 1964 (4) . Sarkisian and Modjtabai 1965 (9) . have given more detailed account on this interesting lesion.

Sex incidence : Seems to be equal in male and female.

Age incidence : 90% of the aneurysmal bone cyst are seen under the age of twenty (7).

Site : It is commonly seen in long bones and spinal column and rarely seen in other bones. When involving long bones it always occurs in metaphysis and never beyond the epiphyseal line (5).

Only four cases so far have been reported in cranio-facial bones (4-7).

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In 1958 Bernier Bhaskar described two cases involving the mandible, one in an eleven years old girl and another in a woman of fifty-nine. These two cases were the second and third cases known to have occurred in the craniofacial skeleton (4). MacCarty gave an account of a case occurring in the occipital bones (7). Our study includes two such cases (Cases I.V.)

CLINICAL MANIFESTATION :

The patient usually gives a history of trauma a short time prior to onset of complaints. Whether trauma has any bearing to the aetiology or just calls attention to the lesion is a matter of much controversy.

When the lesion is in the long bones the patient usually gives a history of pain of a few months duration accompanied by a gradually increasing swelling, and when the swelling is near a joint the pain is likely to be aggravated by movement.

When the lesion is in the vertebral column, the pain and stiffness in the area involved is the first complaint, as the lesion progresses, neurological signs will usually appear. The thoracic lesion may give rise to weakness of the lower limbs and the lumbar lesion may produce disturbance in bowel and bladder control. In cases of advanced lesion the vertebral column collapses causing aggravation of signs.

RADIOLOGY :

The usual radiological appearance is a "Blown out" distension of the periosteum in the diaphysis of a long bone which is limited by the epiphysial plate. It is this appearance that directs attention to the possibility of aneurysmal bone cyst. Distended periosteum is usually outlined by a paper thin subperiosteal bone shell. The bulged out area may present a "soap bubble" appearance.

When the lesion is in the vertebral column and the vertebral body has collapsed the "Blown out" appearance may not be so clear.

In the long bones the lesion is usually eccentric and is limited by the epiphyseal line.

TREATMENT

There is general agreement that the treatment is always surgical and the results are most gratifying.

MATERIAL :

CASE I :

A thirteen years old girl complained of headache six weeks prior to admission, soon after the headache started she noticed a swelling in the left frontal bone, the pain became so severe that she required heavy sedation for its relief.

Patient gave a history of trauma at the age of four at the site of tumor. On examination there was a swelling in the left frontal and parietal bones soft in some areas tense in others, it did not involve the scalp and measured 7 X 12 Cm.

There was no abnormality in central nervous system. Blood pressure was 120/80, chest X ray was normal, Skull X rays showed a cystic mass with a thin wall having blown out appearance and there were trabeculae in the cyst. Cerebral angiography was normal.

Operation :

Under general anesthesia the mass was removed and the exposed dura was covered with skin graft. (Fig 1. 2. 3. 4)

Histology :

The lesion consists of honey-comb spaces, between and on its walls. This is a soft reddish brown tissue. This tissue is permeated with thin walled blood spaces with haemorrhages between them.

These blood vessels have no muscle cell in their walls.

There are also phagocytes laden with haemosiderine and multinuclear giant or endothelial like cells, which line and surround these spaces.

Between vascular spaces and surrounding them is trabeculae of connective and bone tissue.

The giant cells are mostly seen in haemorrhagic areas. In compact and haemorrhagic areas where organization is taking place thrombosis of the small blood vessels can be seen. (Fig. 10. 14. 15. 19.)

CASE II :

A boy of 13 years of age was admitted to the hospital complaining of pain and swelling in the lower region. The swelling was noticed a year prior to admission but pain started a few months later with the swelling increasing in size. For the last month he had noticed weakness in both legs and difficulty with micturition. There was no his-

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tory of trauma. On examination the child appeared wellbuilt. There was a swelling in the lower lumbar region measuring 10 X 8 Cm. in diameters, it was tender, tense and fixed to the deep structure the overlying skin was normal.

X Ray Examination :

X ray examination revealed a cystic swelling with blown out appearance in the spinal process of the third lumbar vertebra limited by a thin layer of bone (Fig. 8 and 9).

A myelogram showed obstruction at the level of the third lumbar vertebra. A provisional diagnosis of aneurysmal bone cyst was made which was confirmed later by biopsy (Fig. 8. 9.).

Operation :

The whole tumor including the spinous processes of second and third vertebra and adjoining tissue was removed and dura at this level was exposed. The patient made an eventful recovery the weakness of the legs disappeared, completely and he gained full control of his micturition.

Histology :

Microscopic examination under low power demonstrates a cellular tissue interlaced with many cavities filled with fresh blood. Under higher power the tissue is composed of two components :

- 1- A well vascularized fibroadipose tissue at the periphery.
- 2- A more compact tissue containing numerous multinucleated giant cells and bony trabeculae.

The latter part borders the large and small cavities. Some of these cavities are lined by a thin endothelium with an underlying reticulum, while others are devoid of endothelial lining. There are many slits and smaller blood channels in the more compact zone. Some of these channels are filled with blood or plasma. Many of the vessels shows a muscle coat with or without intimal fibrous proliferation (Fig. 17. 20.).

There are some arteries and veins whose walls are quite thickened and their lumens are obliterated by the intimal thickening.

Outside of the vascular channels and surrounding the cavities many hemosiderine laden macrophages are present.

CASE III :

A five years old girl was admitted to the hospital complaining of swelling and pain of two months duration in the lower end of the left forearm. There was no history of trauma. On examination there was a swelling over the lower end of the left Ulna which was tender on pressure. The skin was intact and there was no limitation of movement.

X ray examination :

X ray showed a cystic swelling in the lower end of Ulna limited by epiphyseal plate. The periosteum was thickened and there was a thin layer of new bone under the periosteum. The cyst had a typical soap bubble appearance. (Fig. 6. 7.).

Operation :

The cyst was exposed and removed there was profuse bleeding from lesion during the operation but it stopped completely after the cyst was removed.

Histology :

There is a distinct fibrous stroma but fewer giant cells. Blood filled cavities and vessels are of the same architecture as mentioned in case II. There are zones of calcification and intimal fibrous proliferation also.

Case IV.

A twenty years old woman was admitted to the hospital complaining of severe pain of many months durations in the right iliac fossa. The pain started after a full three months prior to her admission and the pain became aggravated by movement.

X ray :

X ray of the iliac bone showed a cystic swelling with a soap bubble appearance. (Fig. 15).

Operation :

The lesion was exposed and curetted out, there was profuse bleeding but the bleeding stopped as soon as the cyst was exposed. The patient made an eventful recovery.

Histology :

There are many dense bony trabeculae embedded in a reactive fibrous stroma.

Numerous giant cells and macrophages are distributed among bony trabecular. Some vascular spaces and irregular cavities are interlacing the bony structure and fibrous tissue.

DISCUSSION

Although it is maintained that trauma has no part in etiology of this tumor (3) two of our cases gave a history of trauma case I four years prior to onset of symptoms and case IV a fall initiated the pain.

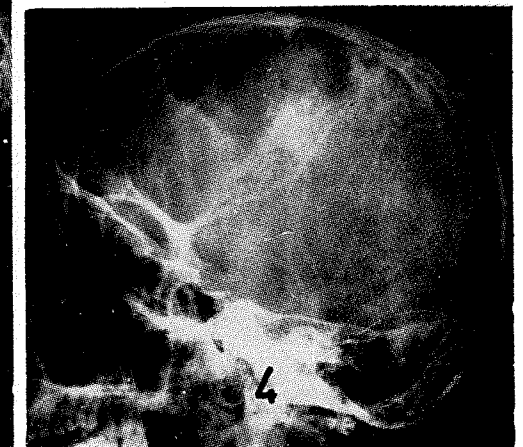
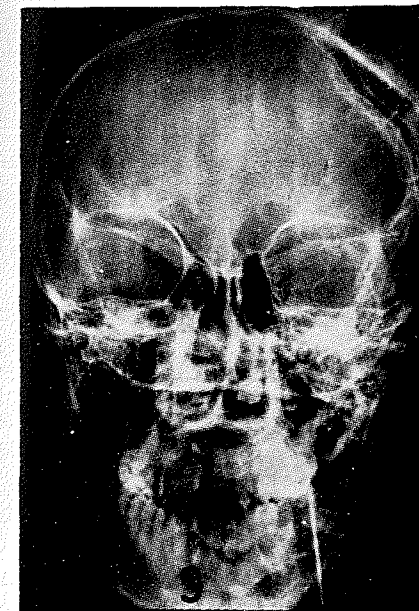
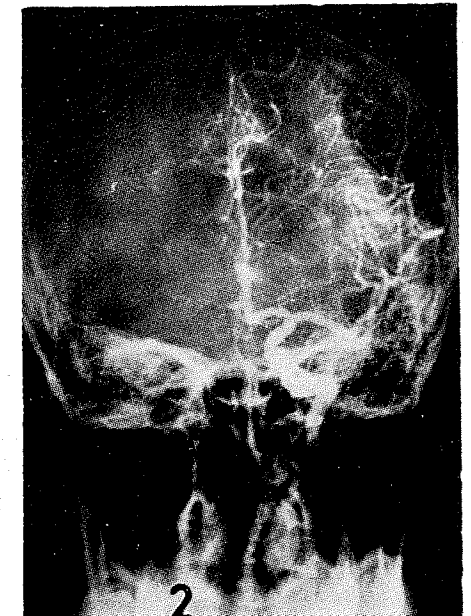
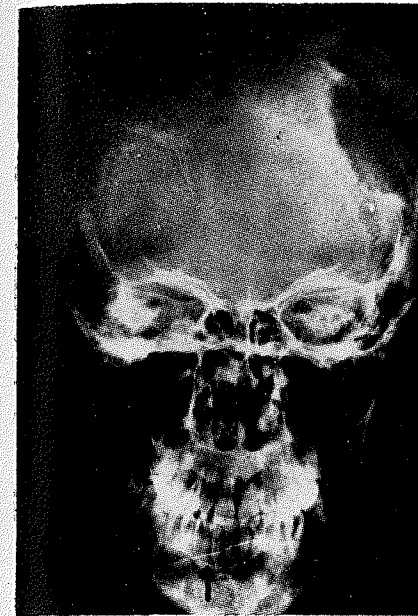
Bleeding in a pre-existing bone lesion is thought to be another etiological factor (6) but in cases where the whole lesion has been excised and studied carefully there has been no evidence of pre-existing pathology (8)

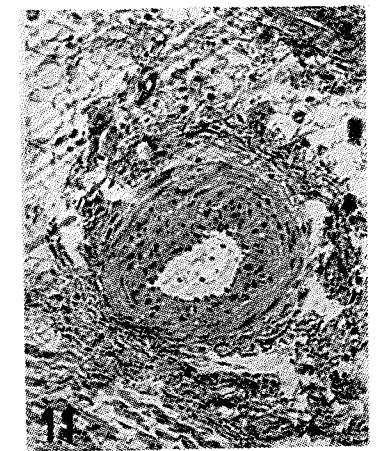
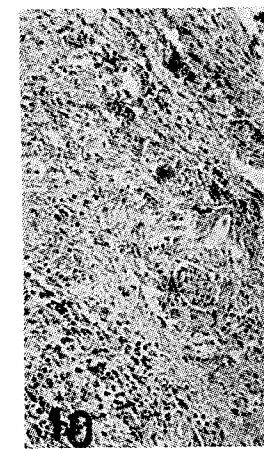
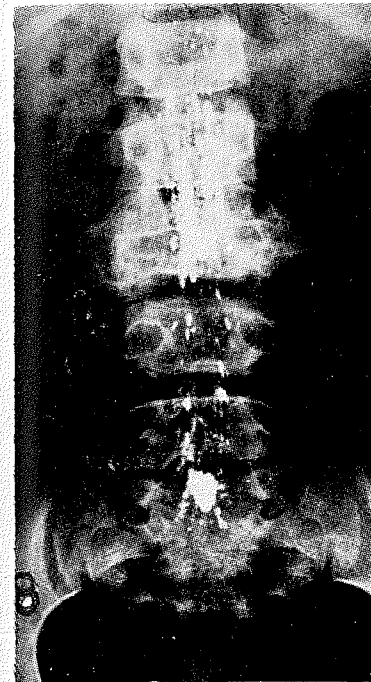
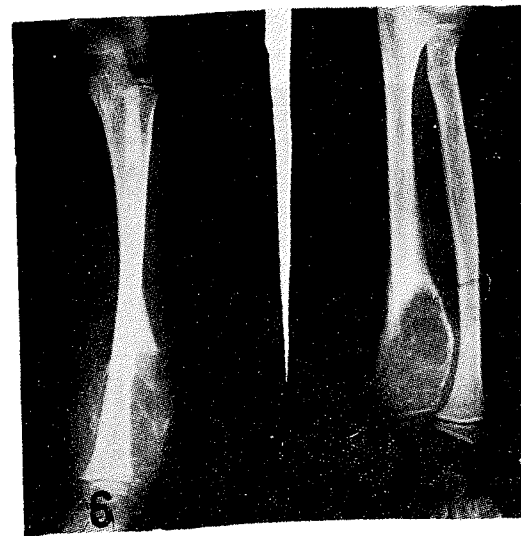
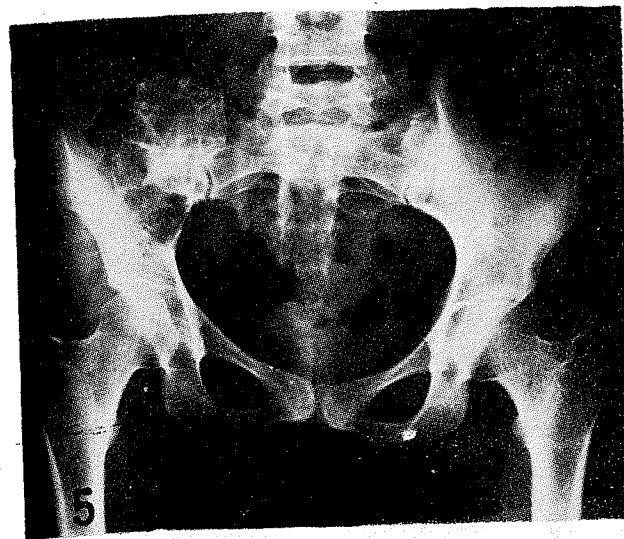
liechtenstein (6) believes the most likely mechanism is something that alters the blood supply of bone producing reabsorption of both cortex and medulla, raising the periosteum, with formation of new bone sub-periosteally and progressive expansion of lesion.

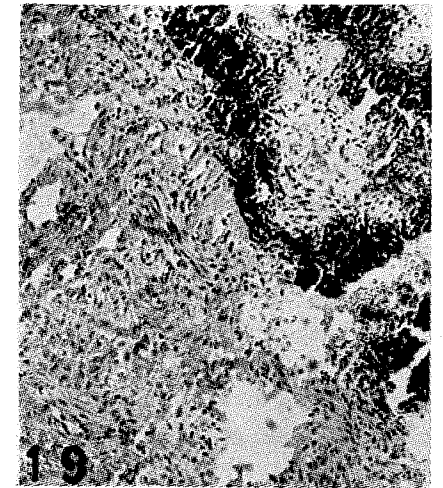
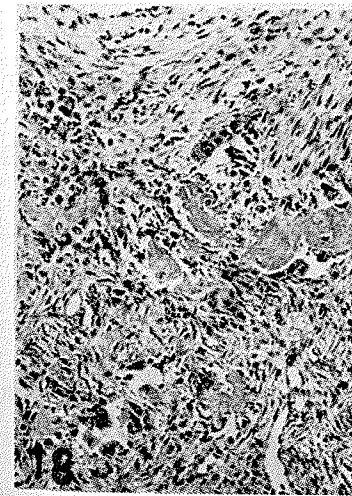
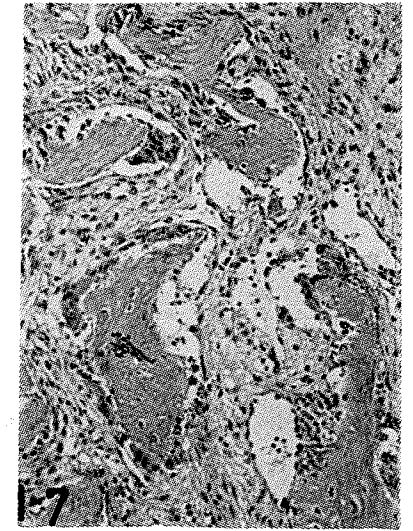
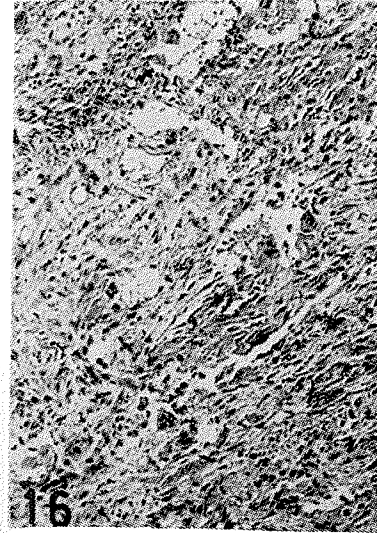
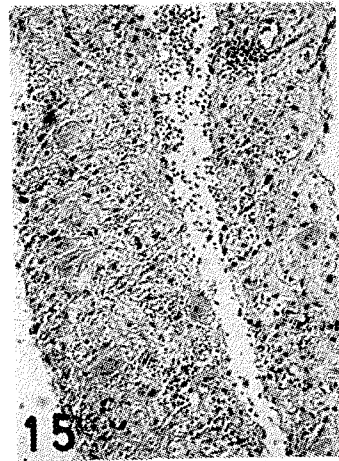
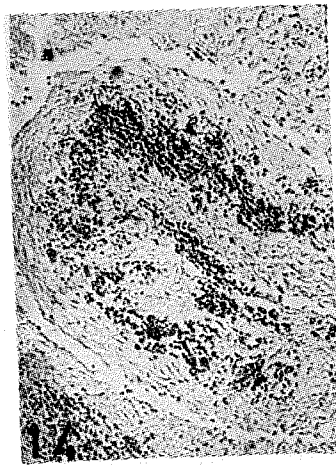
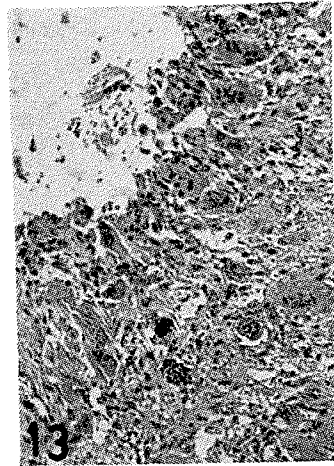
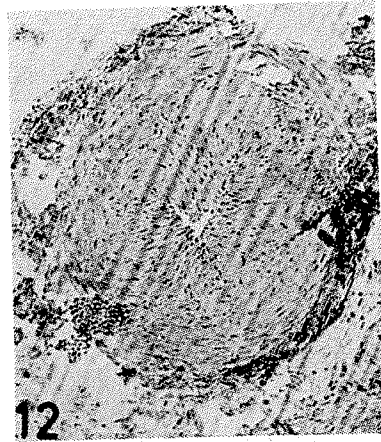
The presence of multinuclear giant cells, specially aggregating around the blood spaces with areas of bleeding, gives the impression of reactionary nature compared with giant cells one encounters in osteoclastoma which is of neoplastic origin.

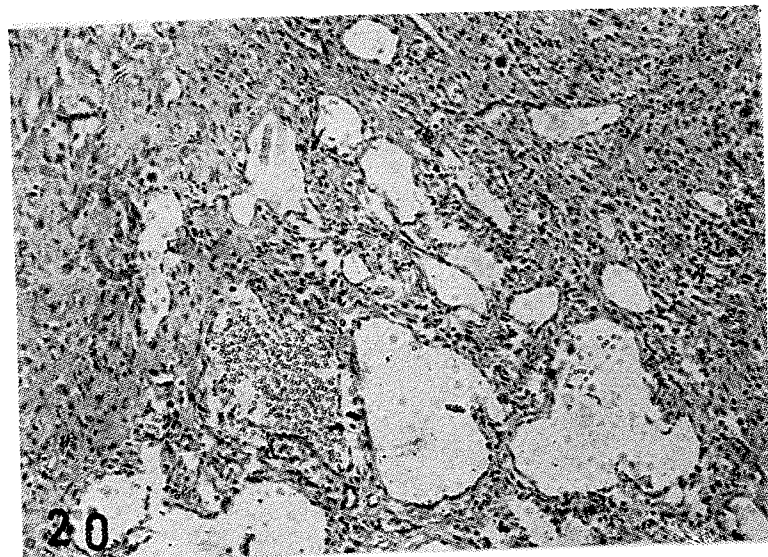
We have come across thrombosis in small blood vessels in two of our cases (9) and it is possible that trauma has caused hemorrhage or thrombosis which have produces an active dynamic process leading to the formation of the lesion.

Edling believes that Aneurysmal Bone Cyst is a manifestation of subperiosteal dysplasia. We do not agree with this, and believe that Aneurysmal Bone Cyst is a separata entity with special pathological, radiological and clinical manifestations.









Legend of figures

- Figs. 1-3-4. Case I. A blown out appearance in left parietal bones.
 Fig. 2. Case I. Normal Cerebral angiography.
 Fig. 5. Case IV. Blown out appearance in right iliac bone.
 Fig. 6-7. Case III. Blown out appearance in lower end of Ulna limited by epiphyseal plate.
 Fig. 8-9. Case II. Myelography, blocked at the level of the third lumbar vertebra.
 Fig. 10. Case I. Thrombosis in a small vessel in state of organization.
 Haemorrhagic area in state of organization.
 Hema. Eosine.
 Fig. 11. Case II. Endofibrosis in a small vessel. Hema. Eosine.
 Fig. 12. Case II. Endofibrosis in a small vessel in state of Organization.
 Fig. 13. Case III. Giant cell abutting on the walls of blood spaces.
 Fig. 14-15. Case I. A blood space with reactionary tissue; the walls of the space do not have normal vessels structure.
 Fig. 16 Case IV. Fibrous tissue with giant cell and hemosiderine pigment.
 Fig. 17-18-19. Case IV. Fibrous tissue with macrophage laden with hemosiderine pigment.
 Fig. 20. Case IV. Fibrous tissue with lined spaces full of red blood cells, stasis in small vessels.

Summary

In this paper four cases of aneurysmal bone cyst are reported and the different etiological factors considered.

We believe that aneurysmal bone cyst disease entity with pathological, radiological and clinical characteristics.

Resumé

Nous avons étudié quatre cas du kyste anévrysmal de l'os et nous avons mentionné les différents facteurs étiologiques probables de cette maladie.

Nous pensons que le kyste anévrysmal de l'os est une entité clinique, pathologique et radiologique.

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