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Contribution to the Histological and Microradiographic Study of the Craniostenosis

By

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With 10 Figures

Summary

The authors describe the microradiographical and histological aspects of ill sutures in cases of trigonocephalies, brachycephalies and oxycephalies. This is based on the study of fragments coming from 30 cases of craniostenosis.

They question the responsability of the dura mater and invoke an encephalic cause producing by error forces at the level of the sutural areas. They also stress the absence of chondroid tissue at this particular level.

Keywords: Craniostenosis; pathogeny.

Introduction

The pathology of sutures interest both the neuro- and the maxillo-facial surgeon due to the close relationship existing between the cranium and the face. This characteristic appears in numerous dysmorphies but mainly in those affecting the orbito-frontal segment [6] in cases of premature closing of the sutures, whether of the cranium or the face.

The cranial and the facial sutures have a similar structure and therefore incite the stomatologist to study cranio-stenosis instead of facio-stenosis which is not so well documented. Indeed, it is known that the small volume of the brain pan in craniostenosis brings about the extrusion of the cerebral contents by the exeresis of osseous fragments, which can be studied by the pathologist, whereas in contrast, the facial exiguity is treated with respect to the skeletal material.

The anatomo-pathological descriptions of the craniostenosis are rare and fragmentary as the exeretic fragments studied come from a parasutural or presumed sutural areas. This explains why the first stages of the stenosis and its development are still little known. For this reason we have been motivated to describe the anatomo-microscopical aspect of the bone of the cranial dome and sutural areas coming from abnormal heads.

This study only covers the problem of simple craniostenosis, *i.e.* cranial deformations generally induced by the premature closing of one single suture in a very specific area.

Material and Methods

1. Material

We observed pathological sutures from 30 cases of craniostenosis. The metopic suture, which results in the deformation of the frontal bone in 11 cases (trigonocephaly), the sagittal suture which stops the development of the parietal bones (scaphocephaly) from 8 children, the coronal suture from 6 children (plagiocephaly and brachycephaly) and, last but not least, various pathological sutures have been studied in 5 cases of oxycephaly. Except for the cases of oxycephalies, the majority of the patients' ages ranged between 5 months and 3 years.

The fragments come from a suture prematurely closed or a parasutural area crossed by a suture. In general, we have studied fragments which come from a presumed sutural area: the metopic area in the trigonocephalies, the sagittal area in the scaphocephalies and the coronal area in the brachycephalies or the plagiocephalies.

To examine the oxycephalies, we took various sutural remaining fragments as well as a spheno-frontal suture.

The choice of the reference material has resulted in practical difficulties. Indeed, the majority of the children whose crania have been studied suffered from renal or cardiac illness with direct or indirect skeletal repercussions. Therefore, we decided to limit our reference to the case of a young girl who suddenly died at 9 months from a thymoma revealed at autopsy.

2. Methods

We have prepared thick not decalcified sections to be used for the microscopical analysis and which can still be stained. To obtain thick sections, the specimens have been embedded in methyl methacrylate according to the conventional laboratory techniques described previously [2] and cut in a series of sections, 150 microns thick, by means of a special diamond saw (Safag, type 32, Bienne, Switzerland).

These sections are then brought to a uniform thickness of 80 microns by using a piece of ground glass kept wet with absolute methyl alcohol. The thickness is measured with a high precision micrometer.

The microradiographs were obtained by placing the sections in contact with a fine grain emulsion (Spectroscopic plate-649-0).

The X-rays are produced by a Machlett tube with a tungsten anode and a 1 mm thick beryllium window, moved by a generator BF 50-20 (Balteau, Liège). The exposure lasts 15 minutes, at 18 mA and 13 kV, for a focal distance of 61 mm. The films are developed for 4 minutes at 20 °C in a D 19 (Kodak) solution, fixed and washed under running water, dried protected from dust and mounted using Canada Balsam as for normal histological preparations.

Table 1

Malformations	Ages	Sex
Trigonocephaly		
1	5 months	male
2	8 months	male
3	10 months	male
4	1 year	female
5	1 year	female
6	1 year	male
7	1 year	male
8	1.5 years	male
9	1.5 years	male
10	4 years	female
11	5 years	male
Scaphocephaly		
12	3 months	male
13	4 months	male
14	5 months	male
15	7 months	male
16	9 months	male
17	2 years	male
18	2 years	male
19	3 years	female
Plagiocephaly		
20	4 months	male
21	2 years, 3 months	male
22	2.5 years	male
Brachycephaly		
23	6 months	female
24	8 months	female
25	7.5 months	male
Oxycephaly		
26	1 year	male
27	2 years, 3 months	male
28	6 years	male
29	9 years	male
30	28 years	male

Results

We present here a series of metopic sutures in the process of obliteration or already closed, in order to observe the phenomena causing the craniostenosis.

We have add an observation of brachycephaly with the same characteristics as the trigonocephaly and will show that the aspect of the oxycephaly cannot be compared with the previous anomalies.

Fig. 1 relates to the reference material. It is a perfectly normal metopic suture taken from a young girl who suddenly died at 9 months. The metopic suture which is wide open and transparent to X-rays is easy to trace. The frontal bones appear on both sides. Both of them already contain two tables, an internal left and right table which mainly consist of lamellar bone and are very similar. The one on the right of the figure contains more canals but this difference is not fundamental. On the other hand, we have to stress the difference existing between both sides regarding the external table. On the left of the figure, we can see long parallel structures obliquely orientated towards the suture and the exterior of the cranium. On the right, the spaces between the osseous trabecular are parallel to the scalp. This suggests that the opposite bones are not submitted to the same forces. A more detailed description would go beyond the limits of this study and will be the topic of a future publication.

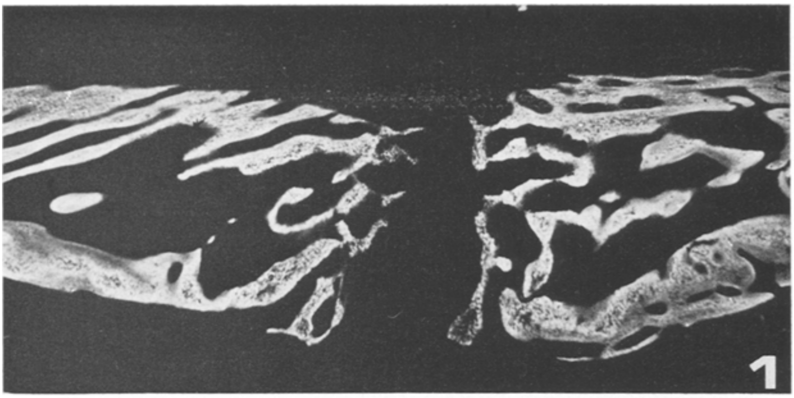
Let us only add that each of the osseous extremities in front countains numerous highly calcified islets which prove the presence of the same "chondroid" tissue as that in the symphyseal area of a mandible in the process of development. This tissue has also been the subject of a recent publication [4].

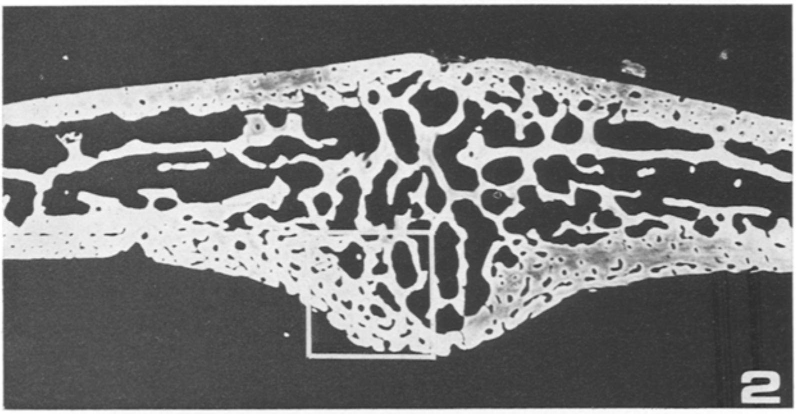
Thanks to this reference figure, we will be able to compare with the observations made in cases of children suffering from trigonocephaly.

The anatomo-pathological aspects of the sutural areas which can be studied mainly depend on the successful tracing of the suture.

Fig. 2 shows the aspect observed at the level of the metopic suture of a 10-month-old child suffering from trigonocephaly. The location of the suture appears very clearly. It corresponds to a very well-marked curve of the internal table and to a little notch in the external table. Moreover, the trabeculae of cancellous tissue which form the diploe are perpendicular to the lamellae, whereas they are parallel in the adjacent bones.

Fig. 3, on the contrary shows the microradiography of a slice of a metopic suture from a 19-month-old child which also suffered from a trigonocephaly; the sutural area cannot be traced anymore. Moreover, we can observe small vascular channels perpendicular to the surface of the external table which diverge as indicated by the arrows. This divergence of the channels suggests that the sutural area appears in the chosen field. However, any further conclusion is impossible. The main point to keep in mind is that Figs. 2 and 3 show completely





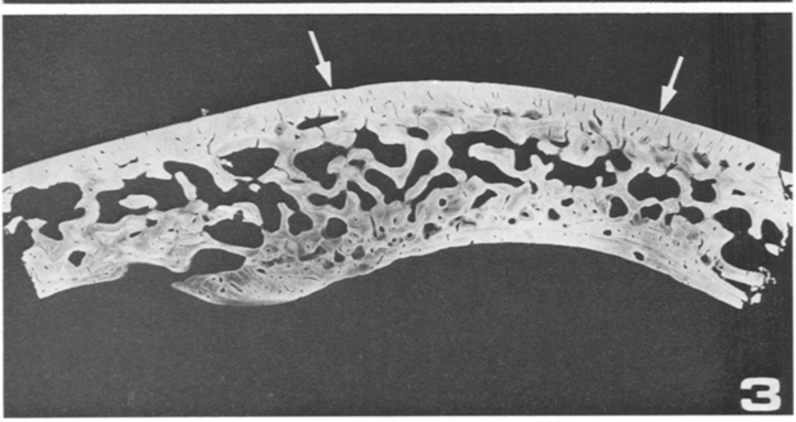


Fig. 1. Microradiographical structure of a metopic suture coming from a 9-month-old girl (×14)

Fig. 2. Microradiographical structure of the metopic area in a 10-month-old child suffering from trigonocephaly (×8). The location of the suture appears very clearly

Fig. 3. Microradiographical structure of comparable fragment coming from a 19-month-old child suffering from trigonocephaly (×4). The sutural area does not appear anymore

different aspects from the ones observed in Fig. 1. Therefore, we must admit that the acquisition of a synostosis is a phenomenon which appears very early in the development of the cranium.

Fig. 3 which results from the study of a sample coming from a 19-month-old child cannot but confirm this interpretation. Indeed, it shows that the visible bony tissue is exclusively lamellar as in adults, with numerous traces of Haversian changes.

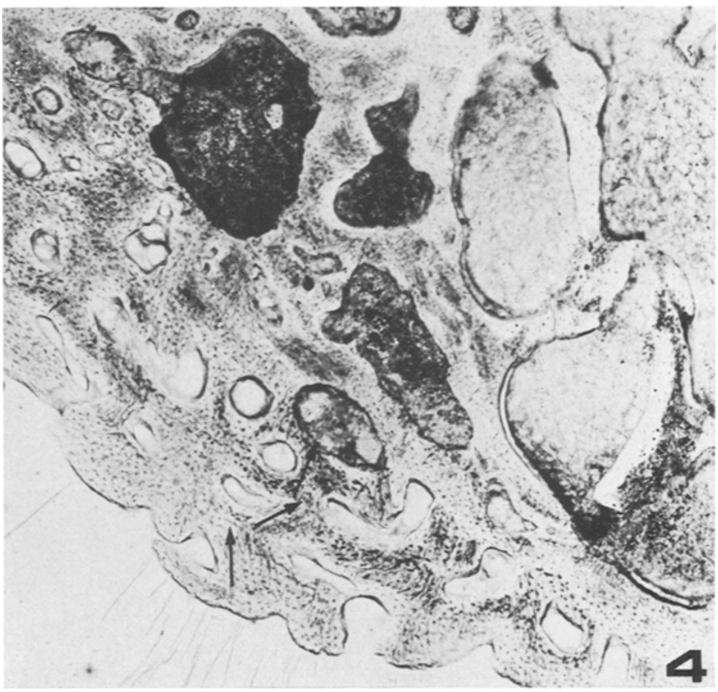


Fig. 4. The field coming from the slice stained with methylene blue corresponds to the area framed in the rectangle in Fig. 2 (×40). The arrows indicate the undefined orientation of the dura-mater fibres

In Fig. 2, the area in the rectangle has been selected in the slice after surface staining by methylene blue and reproduced enlarged in Fig. 4.

The osseous tissue present in this field is mainly reticulated fibrous tissue. It is also clear that the vascular channels are no Haversian systems. The first ones are formed "de novo" whereas the second ones result from an osteoclastic resorption followed by a new bony apposition. This picture shows the presence, proved by slight striation, of dura-mater fibres included in bony tissue. These fibres are characteristic because they have no specific direction. The arrows have been added to this figure to draw the attention of the reader towards this pattern. Sometimes the dura-mater fibres are more or

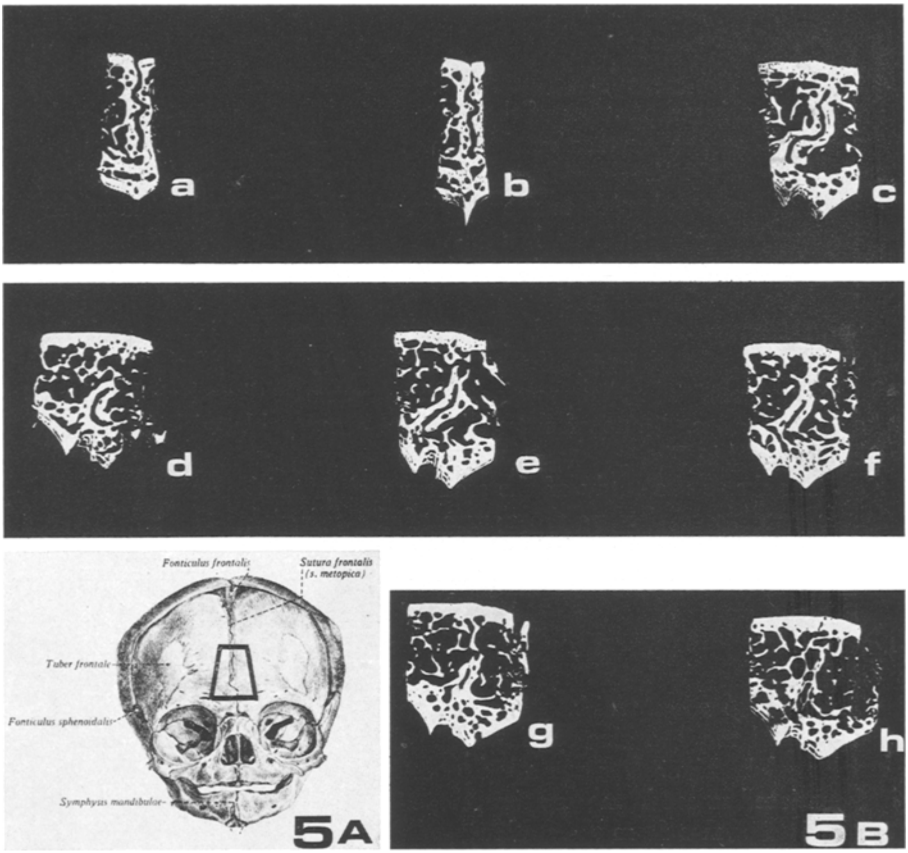


Fig. 5. Study of a metopic suture coming from a 1-year-old child suffering from trigonocephaly. In A, where it comes from; in B, microradiographies of 8 transversal sections designated by letters a to h (×2.5)

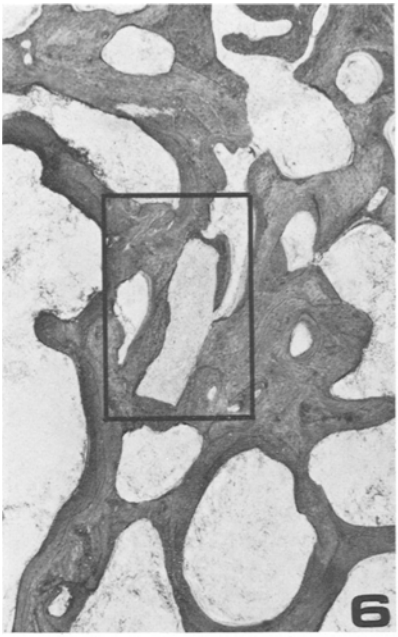
less perpendicular to the surface of the internal table, sometimes they are oblique, whatever the direction. This statement is very interesting. Indeed, it indicates that the dura-mater fibres let themselves be passively included in the ossification. Consequently, they do not guide or give any direction to this ossification. So is it that from this stage we can already question theories currently stated about the process of development of a normal cranium.

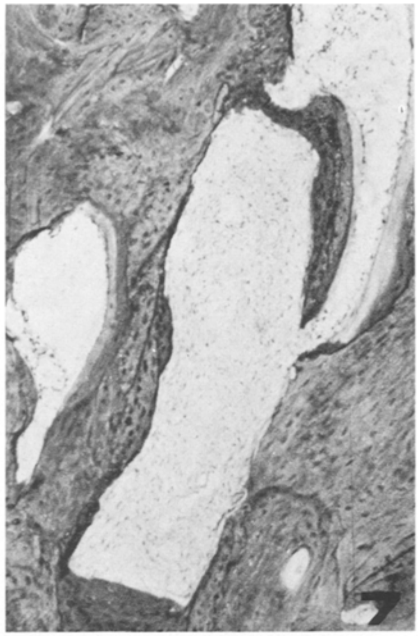
The third case of trigonocephaly concerns an uncompleted synostosis observed in a 1-year-old child. The frame drawn on Fig. 5 shows the place from where the sample was taken. The fragment, 3 cm long, narrower at the vertex than the opposite end, produced transverse sections, 7 mm deep by 6 mm wide. Fig. 5 gathers the microradiographs from eight consecutive sections from the vertex to the fronto-nasal suture. The sections identified by the letters a to h illustrate the remaining traces of the sutures or what amounts to the same thing the extent of the sysnostosis. They are set out in such a way that the exocranial side always faces the superior edge of the figure. In a, the suture appears as a narrow space which meanders from the external table to the proximity of the internal table which is already continuous. In b, the exocranial extremity of the suture is reduced to a small opening and, in c, the sutural space does not continue any further to the external table. Starting in figure d, this progressively reduces and practically disappears in g and h. Simultaneously, the cancellous bone takes the place of the remains of the suture. The disappearing of the suture is directly related to a bony change. None of these slices are comparable with what can be observed in Fig. 1.

Thanks to the study of the sections stained with methylene blue, it is possible to observe the nature of the soft tissue located in the suture as well as the relationship between the dura mater and the sutural area. To illustrate the phenomena at the level of this suture, in Fig. 6 we have reproduced enlarged what was observable in section h in Fig. 5. The bony tissue represented is everywhere lamellar and the links of the diploe are occupied by well-differentiated marrow. The sutural area, which appears as a canal limited by a wall of compact bone, contains another tissue. The framed area reproduced enlarged in Fig. 7 leads us to the conclusion that it is slightly differentiated mesenchymatous tissue.

Fig. 8 comes from the section used for the microradiography of c in Fig. 5. This field shows part of the internal table, nearby the dura-mater, observed after staining of the section on the surface with methylene blue. An osteoid seam, whose low staining distinctly contrasts with that of the bone, is clearly visible. This osteoid seam is itself covered by fusiform cells looking like osteoblasts.

Such an image corresponds without any doubt to an osteogenesis in process which thickens the internal table and equally reduces the volume of the cranium. Moreover, this osteogenesis also results in including the dura-mater fibres in the osseous tissue. The latter appear so clearly in the reference field that it is not necessary to stress it. As in Fig. 4, these fibres are set out perpendicularly to the





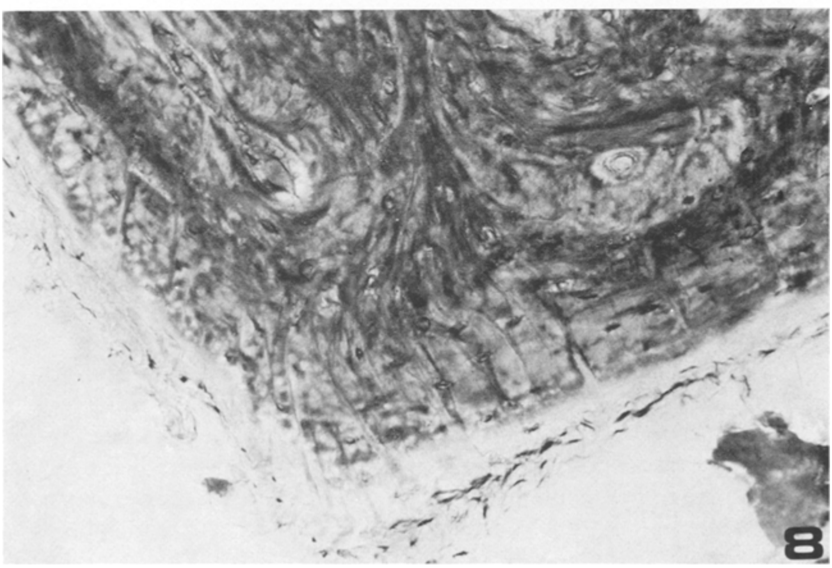


Fig. 6. Slice h, stained with methylene blue and enlarged (×24)

Fig. 7. The rectangle limits the enlarged sutural area (×70)

Fig. 8. Endocranial side of the slice c characterized by an osteogenesis including the dura-mater fibres (×150)

osteogenesis. This demonstrates that the osteogenesis does not develop according to a line oriented by the dura mater.

The other simple craniostenosis presents the same characteristics as trigonocephalies. Fig. 9 is a reproduction of a section through a bony part of the frontoparietal area of an 8-month-old child suffering from brachycephaly. The coronal suture is, in this case, always open and for the normal bone it should be noted that the opposite osseous extremities are not strictly symetrical as the orientation of the bone

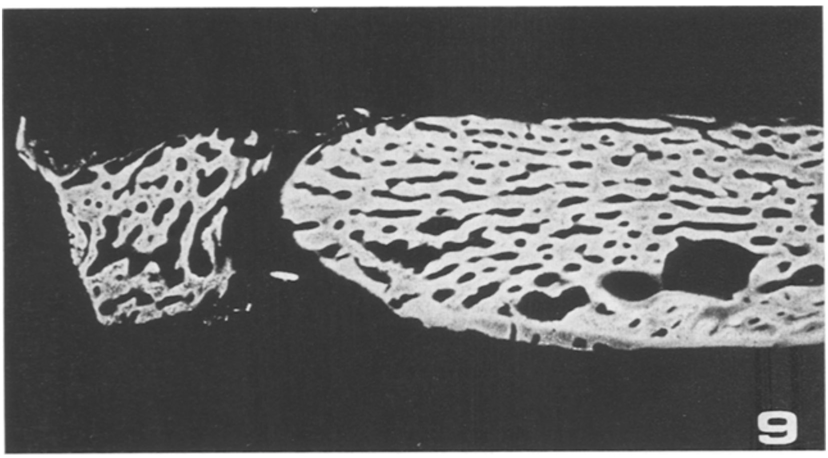


Fig. 9. Microradiographical structure of a slice coming from the osseous region from the fronto-parietal area of a 8-month-old child suffering from brachycephaly (×8)

trabeculae in the osseous tissues varies from one another. As in normal bone (Fig. 1), the chondroid tissue is also visible, but at one extremity only and in very low quantity. This figure does not illustrate exactly the same material as the one found in a normal suture (Fig. 1).

The microradiographical aspect of oxycephaly differs even more from the normal cranium. Fig. 10 represents the microradiographical aspect of a section coming from a 27-month-old child suffering from an oxycephaly. "A" refers to a parietal piece and "B" to the metopic area. The endocranial side is irregular because the internal table is hollowed with a consequent deformation of the diploe. This is made out of cavities smaller than these of the normal diploe. The external table is the centre of a very rapid lamellar deposition. As will be demonstrated in a coming publication, the general aspect of the vault

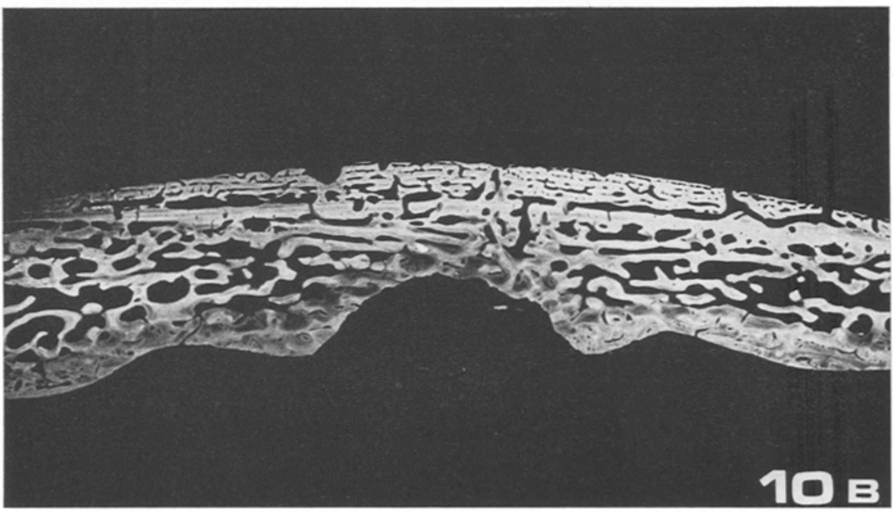


Fig. 10. Microradiographical structure of an oxycephaly affecting a 27-month-old child: In A, the sagittal suture (×5) and, in B the metopic suture prematurely obliterated (×6)

of the cranium in oxycephaly can be mistaken with that of Crouzon's disease.

Nevertheless, the first case being initially older, it leads one to think that the ossification process is slower and that the hollowed notches are not so deep as in the second case.

Discussion

The consequence of the observations presented in this paper is evidently that there are two types of craniostenosis.

In the first group, the premature synostosis is definite and the bony tissue is not normal at all. In the second group, on the contrary the sutural area is occupied by soft tissue and the osseous extremities, in front, have a normal appearance (Fig. 9). How is the difference explainable?

The comparison between Figs. 1 and 2 has to be taken as a reference as it concerns observations from children of the same age. The only difference being that the child suffering from trigonocephaly (Fig. 2) is one month older than the normal one. But, according to what we know about the growth process it clearly appears that such a difference is not sufficient to replace the structure visible in Fig. 1 by that in Fig. 2. Therefore, the difference in age definitely can not explain the structural difference.

The comparison of these two documents leads one to admit that the development mechanism of the cranium in the case of trigonocephaly is completely different from the normal. Nowadays, it is still impossible to state that the error can be attributed only to the mesenchyma used to form the cranium. We can very well imagine that the centre of the anomaly is the encephalon and that its abnormal development explains the pathological development of the cranium [1, 3].

On the contrary, the situation in apparently normal sutural areas is completely different than that shown in Fig. 9. It would neither be the cause of any primary difficulties, nor secondary ones. Nevertheless, maybe surgical operation would prevent its development. In both the normal child (Fig. 1) or in the child suffering from brachycephaly (Fig. 9), we observed an asymmetry of the opposite osseous edges. This is not really surprising. Indeed, as everyone knows, the reference anatomical image of the sutures present at all levels filling areas where, in the case of the identation of a bone, it automatically induces a groove in front. Moreover, the left side of Fig. 1 clearly shows that the bony trabeculae which constitute the external table are obliquely orientated towards the suture of the cranial surface. Such an aspect suggests that the osteogenesis is the result of mechanical forces such as traction and that these partially explain the development mechanism of the cranium.

The relationship between the mechanical forces on the periosteum drawn by the growing cartilage and the development mechanism of the bony trabeculae, at the origin of the diaphysar symptom, have been analysed in the study by Lacroix [5] since 1949. On both sides of a neutral point where the tractions counterbalance each other, the trabeculae of the shaft are opposed giving to the diaphysis the aspects of 2 pine-cones joined at their bases. At the level of the neutral point, the bone trabeculae are perpendicular to the longitudinal axis of the bone and the neutral point is always located nearer to the less fertile cartilage than to the other.

Due to this observation, we could assume that the premature acquisition of craniostenosis results from abnormal mechanical forces. In this case, the cranial anomaly is the result of an encephalic anomaly which is the only thing able to cause the abnormal mechanical forces.

The existence of mechanical forces at the level of the suture is also proved, according to us, by the presence of chondroid tissue. This tissue, observed for the first time by Shaffer in 1888 [8], has also been mentioned by Goret-Nicaise and Dhem in 1982. They noticed its permanent presence in the foetal mandibular symphysis. At this level also, there is traction related to the tongue development and the existence of tissue also showed histologically the results of mechanical traction.

This automatically leads us to consider the role played by the pachymeninx in the acquisition of a craniostenosis. According to the work of Moss [7] the development of the vault of the cranium is the harmonious result of a pachymeninx correctly settled. On the contrary, the premature acquisition of a craniostenosis would result in an abnormal implantation of the pachymeninx at the basis of the cranium.

Actually, such an interpretation we consider to be incompatible with the observations mentioned above. Indeed, as described, the influence of the mechanical forces clearly appears on the external table of the bone and not on the table in contact with the pachymeninx. Moreover, as appears in Fig. 4, the implantation of the dura-mater fibres in the osseous tissue takes place without any specific order. It is not rare to observe in the same microscopical field various groups of differently oriented fibres. Moreover, in Fig. 8, there is a proven osteogenesic area shown by a preosseous edge covered by osteoblasts including dura-mater fibres perpendicularly oriented to it. In any case of osteogenesis it is guided by soft tissues, which follows a direction parallel to the included fibres. A remarkable example has been given by Dhem and Vincent [2] regarding the incorporation of a patellar ligament in the tibial tuberosity of a growing dog.

On basis of both Figs. 4 and 8, we can state that the dura-mater fibres do not play any role in the development of the osseous tissue. The structures studied have to be compared with the situation observed in which the periodontal ligament has been implanted.

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