Retinal elements in an amorphous twin

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Amorphous twins are rare and the case reported here showed embryonic retinal tissue including the formation of rosettes. This appears to be the first case of such a twin showing retinal structures. The specimen was received in the laboratory as an “ovoid mass” attached to the placenta of a normal healthy male child born of a woman with two previous full-term normal deliveries.

Investigations

MACROSCOPICAL
The specimen consisted of a placenta with the attached portion of the cord of the normal foetus towards one pole. The umbilical vessels of this cord traversed the placenta and were continuous with another shrivelled-up cord 6 cm. long hooded by a globular mass 6–8 cm. in diameter clothed with skin containing sparse hair. No definite structures were made out externally (Fig. 1, overleaf). The cut surface showed a central cavity 3 cm. in diameter having dark brown tags surrounded by firm brown tissue with tiny glistening areas simulating cartilage with adjoining chips of bone.

MICROSCOPICAL
The skin showed well-developed stratified squamous epithelium and adnexa. The mass consisted essentially of loose connective tissue showing pronounced oedema. There were widely scattered numerous tiny blood vessels and endothelial lined spaces, possibly dilated lymphatics. A predominant additional feature was the richly vascularized foetal fat and strands of muscle fibres with characteristic transverse striations. Focal collections of sympathetic ganglion cells, in places forming large groups, were also seen. Individual ganglion cells were surrounded by a single or double layer of monocytoid cells, mostly resembling lymphocytes. In one group there was continuity of nerve fibres with the ganglionic mass. Several well-developed nerve fibres were also distributed irregularly throughout the specimen.

In sections taken from the central cystic area the histological features were of great interest. The cavity wall was formed of loose connective tissue together with numerous blood-filled cavernous spaces and dilated channels. The space was partially but clearly lined by a single layer of cuboidal, flattened, or low columnar epithelium, in parts pseudo-stratified, resting on loose connective tissue. Some of these cells showed intracytoplasmic dark-brownish pigment. The close proximity in places of this lining to the overlying skin suggested a probable anatomical inter-relationship between the two, but no direct continuity could be traced in the sections examined.

The most striking and characteristic feature was the presence of retinal elements which were arranged haphazardly, though mostly disposed around folded spaces or occurring as sparse islands.
within loose and oedematous connective tissue. Nucleated cells and cells resembling rods and cones with distinct limiting membranes and prominent fibrils in direct continuity with these nucleated cells gave striking semblance to structures normally seen in a well-formed retina (Fig. 2).

More characteristic still were the rosettes that had formed amidst them closely simulating the rosettes commonly encountered in retinoblastomata (Figs 3 and 4, opposite). The rosettes were also surrounded by fibrils. Besides these retinal elements, a structure resembling the optic cup of the embryonic eye was seen lying free within mixed connective tissue.
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**FIG. 3** Rosettes under higher magnification, showing characteristic limiting membrane. These structures resemble those commonly seen in retinoblastoma. Haematoxylin and eosin. ×275

**FIG. 4** Same area as Fig. 3. A well-developed rosette with characteristic fibrillary and reticular structures. Phosphotungstic acid haematoxylin stain. ×275

**Comment**

Reviewing and summarizing the nature of this rare condition, Willis (1962) regarded it as a type of acardiac foetus. Amorphi differ greatly in their internal structure but are always clothed with skin. In the present case the microscopic structures included almost all the elements that are known to occur in amorphi. The exceptional finding was the well-defined retinal structure with rosettes similar to those encountered in cases of retinoblastoma. The appearance of rosettes in these two different situations and the finding of rosettes in cases of retinal dysplasia, regarded by Hogan and Zimmerman (1962) as a developmental aberration, may form a part of a hitherto undefined embryological defect.
Retinal tissue culture (Strangeways and Fell, 1926—cited by Willis, 1960) may assist the interpretation of such aberrations.

Summary

A rare case of amorphous twin yielded unusual embryonic retinal elements, including the formation of rosettes similar to those found in retinoblastoma.

Our thanks are due to Dr. S. Janki for referring the specimen to us.

References


