Conducting system in foamy myocardial transformation of infancy

We were interested to read of five cases of foamy myocardial transformation of infancy reported by Suarez et al. The observation of familial occurrence of this rare disease, which often presents with dysrhythmias, is an important one. We have recently had the opportunity to study the conducting system in the heart of an 18 month old boy with foamy myocardial transformation who died suddenly with no antecedent history.

The heart was of normal size and no macroscopic abnormalities were noted. Microscopic infiltrates of foamy myocytes were present within the left ventricle. A large plaque of these cells was closely applied to the connective tissue sheath of the left bundle branch. An abnormal potential conducting pathway was shown, bridging between the origin of the left bundle and the crest of the ventricular septum. This pathway consisted of foamy myocytes and could have acted as the route for a re-entry dysrhythmia (figure).

The examination of the conducting system in childhood is time consuming but not difficult. The technique generates considerably fewer sections than when an adult heart is examined. Foamy myocardial transformation has been rarely diagnosed and treated in life, but screening of siblings may now yield presymptomatic cases. The future management of this condition can but be assisted by as much detailed histopathology as is possible from necropsy specimens.

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References

Figure Bridge of foamy myocytes (arrow) links left bundle (LB) to crest of ventricular septum (VS). (Haematoxylin and eosin.)