

ABSTRACT

Background:

Hirschsprung's disease (HD) is a common cause of intestinal obstruction in children. Full thickness biopsy or resection of involved intestinal segment and its histopathological examination (H&E) is necessary for diagnosis. Ancillary diagnostic tests are required to improve the efficiency of H&E diagnosis. This study evaluates, whether Calretinin immunohistochemistry (IHC), could improve the accuracy and precision in diagnosing HD in clinically suspected cases.

Materials and methods:

Full thickness biopsy or resection specimens of 30 clinically suspected cases of HD were studied. Samples from ganglionic and aganglionic segments for each patient was analysed for presence of ganglion cells and nerve bundle hypertrophy in all the bowel layers using H&E. Similarly, IHC was used to assess ganglion cells and nerve immunoreactivity. Both H&E and IHC assessment was done by two pathologists, one experienced and another in-training pathologist, independently, blinded from each other.

Results:

50% patients were neonates. 87% had short segment involvement. 27% were resected specimens. Five patients were diagnosed as non-HD. Of the

remaining 25 patients, suspicious diagnosis was reported in four patients by Observer1 and one patient by Observer2. Both pathologists could provide definitive diagnosis using IHC for all cases, improving the diagnostic sensitivity by 13.3%. There were 13.8% equivocal observations for ganglion cells with H&E and none with IHC. Interobserver assessment showed statistically significant improvement in Cohen's kappa from 0.574 (moderate agreement) for H&E to Kappa=1 for IHC ($p<0.05$).

Conclusion:

Calretinin IHC is a useful adjunct to H&E in diagnosis of HD, improving the diagnostic sensitivity (diagnose more cases), accuracy (confirm suspicious cases) and precision (reduce interobserver variation). It might be a very useful marker in improving diagnostic accuracy for pathologists with lesser experience. The additional cost of IHC could be justified by improved sensitivity, which would save patient from morbidity and expenses of second look surgery.

Key words:

Hirschsprung's Disease, Calretinin, Immunohistochemistry, Histopathology, Inter Observer Concordance.