Histoid Hansens – The Current Perspective

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Abstract: Histoid leprosy, once thought to occur in patients on dapsonemonotherapy is a well recognised entity also occurring in patients after multidrug therapy, as well as denovo. Since it resembles many conditions like dermatofibromas, neurofibromas, molluscumcontagiosum, sarcoidosis etc., it is likely to be missed out and hence forms the reservoir of infection. Here we report 5 cases of denovohistoid leprosy out of whom 2 patients developed type 2 reaction. The diagnosis was confirmed by slit skin smears and histopathological examination.

Keywords: denovo, histoid leprosy, spindle shaped cells, type 2 reaction.

I. Introduction

Histoid leprosy is a form of multibacillaryhansens characterised by cutaneous and subcutaneous nodules and/or plaques over apparently normal skin with unique histopathological and bacteriological findings. It was first described by Wade1. The incidence varies from 1-2% 2. It can occur after dapsonemonotherapy, after multidrug therapy or denovo. The characteristic histopathological findings are histiocytes packed with numerous acid fast bacilli arranged in whorls or storiform pattern. A thorough knowledge and early detection is needed for this uncommon variant of leprosy which can pose a threat to our elimination drive.

II. Case Reports

Case 1: A 60 year old male patient presented with smooth dome shaped papules and nodules over trunk and extremities of 6 months duration (Fig 1). Bilateral ulnar, radial cutaneous and lateral popliteal nerves were enlarged. Glove and stocking anaesthesia was present. Ear lobe infiltration was present (Fig 2).Madarosis was present. Slit skin smear(SSS) showed a bacteriological index (BI) of 5+.Histopathological examination(HPE) showed thinned out epidermis with storiform pattern of histiocytes and FiteFaraco stain showed heavy bacillary load (Fig 3&4).

Case 2: A 35 year old male patient presented with multiple skin coloured nodules on the trunk and upper extremities of 8 months duration (Fig 5). Bilateral ulnar, radial cutaneous and lateral popliteal nerves were enlarged. Sensations to touch and temperature were decreased over lower parts of both legs and dorsum of feet. Ear lobe infiltration was present. SSS showed BI of 5+ (Fig 6). HPE showed spindle shaped cells arranged in storiform pattern and some vacuolated cells. 2 months after initiation of multidrug therapy (MBMDT) he developed red painful skin lesions with fever and joint pains. Histopathology from the lesions was consistent with erythema nodosumleprosum (ENL). The patient had no recurrence of ENL after the first episode.

Case 3: A 34 year old male patient presented with skin coloured papules and nodules over trunk and upper limbs of 5 months duration and tender nodules of 3 days duration (Fig 7). Bilateral ulnar and radial cutaneous nerves were enlarged and tender. There was no sensory loss. Ear lobe infiltration was present (Fig 8). SSS showed BI of 4+. HPE from skin coloured papule and tender erythematous nodule was consistent with histoidhansens and ENL respectively. The patient was later lost to follow up.

Case 4: A 40 year old male patient presented with multiple skin coloured to erythematous papules and nodules (Fig 9&10) of 3 years duration which was misdiagnosed as allergic contact dermatitis, sarcoidosissete and was treated with steroids before he presented to us. Bilateral enlargement of ulnar nerves was present. There was no sensory loss or ear lobe infiltration. SSS showed a BI of 6+. Biopsy was consistent with histoidhansens.

Case 5: A 45 year old male patient presented with multiple skin coloured papules and nodules over trunk, face, buttocks and extremities of 4 months duration (Fig 11). Bilateral ulnar, radial cutaneous and lateral popliteal nerves were enlarged. Ear lobe infiltration was present. Glove and stocking anaesthesia was present. SSS showed BI of 4+. HPE was consistent with histoidhansens (Fig 12).
There was no prior history of dapsonemonotherapy or multidrug therapy (MDT) in any of the patients. All the patients were started on MBMDT and those with ENL reaction were treated with tapering doses of steroids. One patient was lost to follow up.

III. Discussion

Histoidhansens is considered by some as a variant of lepromatous leprosy. However according to Sehgal, histoid is a distinct form of leprosy and should not always be considered as a variant of LL hansen's. The incidence of histoidhansens in various studies in India ranged from 2.79 - 3.60%. As suggested by Palit and Inamadar, histoid form could serve as a reservoir of leprosy and as a source of new cases.

The 5 cases of histoidhansens in our study were seen over a period of 18 months. All the cases were males with an age range of 34 - 60 years. Sehgal and Srivastava reported that histoid leprosy is more frequent in males than females. There was no prior history of either dapsonemonotherapy or MDT in any of the patients. In a study by G Kalla et al 17 cases out of 25histoidhansens (68%) presented denovo while it was 75% in a study by Bhutani et al. The number of cases of histoidhansens presenting denovo seems to be on the rise as reported in various studies. The number of lesions ranged from 20 to more than 50 in our study. Ear lobule infiltration was seen in 4 patients and madarosis in a single patient. A noticeable feature in some patients is the persistence of the eyebrows. Trunk and upperlimbs were the common sites affected while facial lesions were seen in only 1 patient. The lesions are usually located on the back, buttocks, face, extremities and over the bony prominences, especially around the elbows and knees. Palms and soles are usually unaffected in histoid hansen's. Ulnar nerve was commonly enlarged in all the patients which was in concordance with a study by G Kalla et al. ENL which is supposedly uncommon in histoidhansens was seen in 2 cases, with onset after 2 months of MDT in one patient and prior to MDT in the other. In a study by Mendiratta et al 27% had an episode of ENL while Kaur et al reported 40% of reaction rate among histoid hansen's. BI ranged from 4+ to 6+. Histopathological examination in all the cases showed spindle shaped cells arranged in storiform pattern with positive fitefaraco stain.

IV. Figures

Figure 1: smooth dome shaped papules and nodules over back and upperlimbs

Figure 2: Papules and nodules over ear lobe
Figure 3: H&E 40x showing spindle shaped cells in whorls

Figure 4: FiteFaraco stain showing loads of AFB

Figure 5: Histoid lesions and ENL lesions on the trunk
Figure 6: Slit skin smear showing BI of 5+

Figure 7: Histoid nodules with central umbilication and ENL lesions on the trunk

Figure 8: Bilateral ear lobule infiltration
Figure 9: Erythematous papules and dome shaped nodules over back

Figure 10: Erythematous papules over left upper limb

Figure 11: Histoid lesions over face, trunk and buttocks
Figure 12: H&E 40X showing whorls of spindle shaped cells with positive Fite-Faraco staining

V. Conclusion

Our observation defy the earlier established cause of histoid hansens and is more in concordance with the recent proposition of histoid hansens arising de novo. It can simulate many other conditions thereby leading to misdiagnosis and hence forming a potential source of infection in the community. Presentation of 5 cases in a short span of 18 months signifies an increase in its incidence requiring larger studies over long duration to confirm this observation.

References