



網站：<http://www.taiwanheadache.com.tw/>

頭痛電子報 第149期

發行人：台灣頭痛學會

發刊日期：民國 105 年

12 月

【本期內容】

偏頭痛性小丑症候群

2

國際頭痛學會 Education Committee List of Educational Activities for 2016

6

各位頭痛學會的會員大家好：

頭痛電子報又要來增長大家的知識了。本期刊出由義大癌治療醫院神經科陳建志醫師投稿的「偏頭痛性小丑症候群」。小丑症候群是一種罕見的顱部自律神經障礙，病人在受熱或運動過後，半側顏面會潮紅、出汗。這種病人有可能合併偏頭痛，且需與偏頭痛引起的顱部自律神經症狀鑑別診斷。此症候群雖然令人困擾，幸好只需保守治療。主編雖然從沒遇過這種病人，但 Dr. Goadsby 在 2012 年的 JNNP 曾一口氣提出三個有趣的個案，讓我們藉本文一探究竟！

電子報主編：臺北榮總 陳韋達醫師

秘書處報告

1. 國際頭痛學會 Education Committee List of Educational Activities for 2016，詳情請參閱 p.5

台灣頭痛學會 敬啟

偏頭痛性小丑症候群

元景耳鼻喉科神經科診所 高雄榮民總醫院耳鼻喉頭頸部暈眩門診 義大癌治療醫院神經科

陳建志

小丑症候群 (harlequin syndrome) 是一種相當罕見的顱部自律神經障礙，最早由Drummond和Lance於1988年提出¹，他們描述了5位患者，在接受熱刺激或運動過後，半側顏面會呈現潮紅和出汗，因此命名為小丑症候群。此症候群確切的病因目前仍未知，普遍認為是一側顏面之交感神經功能發生異常所致，有可能會發生於腦幹梗塞、頸動脈剝離、上縱膈腔神經瘤、接受內頸靜脈導管術或胸腔內視鏡交感神經截斷手術後之患者²⁻⁴。

根據神經解剖學，頭部的交感神經支配起自第1~3胸段脊髓，交感神經節前神經元穿過中、下(星狀神經節)頸段交感神經節，在上頸段神經節處與節後神經元發生突觸接合，接著節後神經元會沿著內頸動脈與外頸動脈支配頭部的交感神經功能。倘若沿著內頸動脈的節後神經元因故發生興奮時，會使額頭和眼部之顏面皮膚血管擴張，進而出現潮紅；倘若是沿著外頸動脈的節後神經元因故發生興奮時，則會使顴骨部和上下頷部的顏面皮膚發生潮紅^{3,6,7}，造成小丑症候群。另一可能的成因，是三叉-自律神經 (trigemino-autonomic) 系統的反射異常，引起第3頸段脊髓神經的逆行性放電 (antidromic discharge)，釋放出血管擴張性勝肽，使相對應區域之皮膚血管發生擴張，而環境溫度的改變正好降低了這種跨形式刺激反應的閾值⁷，進而導致小丑症候群。

根據 Viana 等之個案報告 (3 例) 以及文獻回顧 (6 例)⁸，若患者同時具有小丑症候群和頭痛時，該頭痛大多是偏頭痛，因為偏頭痛也是一種自律神經失調的疾病，但總共只有 2 例個案 (Viana 報告中之個案 2 及 3) 是在偏頭痛 (無預兆偏頭痛) 發作時同時併發小丑症候群，可見偏頭痛發作同時併發小丑症候群誠屬罕見。倘若是偏頭痛發作同時併發小丑症候群時，推測是不明原因造成一側顏面部位的自律神經

失調，在每次偏頭痛發作時，僅使對側外頸動脈的交感神經節後神經元發生興奮，進而發生對側顏面潮紅，造成小丑症候群。

要治療偏頭痛併發小丑症候群，需視臨床發作的情形而定。若單純是小丑症候群本身帶來的半側顏面潮紅出汗，建議保守觀察即可。若有明顯的偏頭痛顱部自律神經症狀（兩側結膜紅腫、流淚、鼻塞、流鼻水、眼瞼水腫、前額和顏面流汗）⁹，可以投予消炎止痛藥、抗組織胺或副腎皮質素。長期而言，偏頭痛併發小丑症候群只需保守治療即可，並不需要接受交感神經截斷術，但對病患解說此現象的成因以化解其憂慮是必要的。當然，若要預防該症候群復發，就必須從預防偏頭痛著手，建議投予偏頭痛的標準預防藥物。

參考文獻

1. Lance JW, et al. Harlequin syndrome: the sudden onset of unilateral flushing and sweating. *J Neurol Neurosurg Psychiatry* 1988;51:635-642.
2. Sarikaya H, et al. Harlequin syndrome in spontaneous dissection of the cervical carotid artery. *Neurology* 2008;71:1459.
3. Moon SY, et al. Harlequin syndrome with crossed sympathetic deficit of the face and arm. *J Korean Med Sci* 2005;20:329-330.
4. Swan MC, et al. Iatrogenic harlequin syndrome. *Postgrad Med J* 2003;79:278.
5. Turco GR, et al. Postoperative autonomic deficit: a case of harlequin syndrome. *Anesthesiology* 1996; 85: 1197-1199.
6. Burlacu CL, et al. Coexisting harlequin and Horner syndromes after high thoracic paravertebral anaesthesia. *Br J Anaesth* 2005; 95: 822-824.
7. Culp WJ, et al. Heat and mechanical hyperalgesia induced by capsaicin. Cross modality threshold modulation in human C nociceptors. *Brain* 1989;112:1317-1331.
8. Viana M, et al. Headache in three new cases of Harlequin syndrome with accompanying pharmacological comparison with migraine. *J Neurol Neurosurg Psychiatry* 2012; 83: 663-665.
9. Lai TH, et al. Cranial autonomic symptoms in migraine: characteristics and comparison with cluster headache. *J Neurol Neurosurg Psychiatry* 2009;80:1116-1119.

國際頭痛學會 List of Educational activities for 2016

(further details may also be found at the [IHS webpage](#))

1. The International Headache Academy

The International Headache Academy (iHEAD – IHA in North America) was established to provide a platform for developing the next generation of headache specialists. The programme of the academy includes plenary sessions, as well as practical workshops and interactive masterclasses. Features of the academy include presentations and debate on hot topics in headache management, as well as practical masterclasses. A key feature of the academies is practical, hands-on learning; workshop sessions are focused on developing leadership skills, including setting up clinical trials, presentation skills and publishing work. Delegates of the academies, comprising the next generation of headache specialists from the region, are carefully selected by the Scientific Steering Committee collaborating with the IHS Juniors Committee and regional National Societies.

2. IHS Headache Trainee Program 2016

This project aims at funding physicians from developing countries or countries in transition to visit a specialized headache institution abroad for 8-12 weeks and actively increase knowledge on headache disorders and specialized headache management. It is also intended to spread this knowledge to their home countries without headache infrastructure or expertise. After a clinical training period of up to 12 weeks at a headache centre abroad the award holder is encouraged to return to his/her country of origin in order to apply the acquired knowledge, thus contributing to the advance of patient management locally. One Headache Trainee will be funded for 2016.

3. IHS Visiting Professor Program 2016

Visiting Professors is an initiative of the IHS Education committee with the aim to send headache specialists as representatives of IHS to attend regional meetings, or teach at a headache centre, in countries that might need or want increased headache education and motivation and where, without financial support, attendance of an international specialist would not be possible. IHS will fund two headache specialists (one senior, one junior) to teach at a national or regional meeting/congress. The host country can select a topic and suggest at least three specialists who will fit in with their program. For 2016, visiting professors for up to 4 regional meetings will be supported.

4. The Headache Master Schools.

These have taken place in Brazil, Japan, India and recently in China. They usually consist of learning modules on line before the meeting, a pretest of knowledge, an onsite program with an IHS and local faculty and attendees from the sponsoring countries, and a post test. Proposals come from countries that want these schools and must include a budget and interaction with IHS Board and the Education Committee. A further school is proposed for Latin America in 2017.

5. The IHS Learning center

Located within the IHS website, the [Learning Center](#) contains useful services, such as

access to the Slide Library, containing an expanding collection of downloadable slide decks contributed by major headache specialists worldwide, to the Reading Library, that allows access for reading or downloading books and chapters written by eminent headache specialists worldwide, the Poster Library containing pdfs of posters presented at previous IHCs, and to the IHS Core Curriculum for Neurologists.

6. New Educational Opportunities

At a retreat of the Board it was suggested we find some educational offering for our members in the off year from our biannual congresses. Dr. Purdy in conjunction with Dr. Dodick proposed to the board that a course on secondary headaches be held at the European Headache and Migraine Trust International Congress (EHMITC) in Glasgow Scotland this September. Working in conjunction with the British Association for the Study of Headache (BASH), a course has been approved by the SPC of EHMITC. This one-day course will consist of cases and interaction around secondary headaches, which we hope will satisfy repeated request from our members for such a course. The faculty for this course will consist of members of the IHS and BASH.

本電子報以電子郵件方式寄發內容包括台灣頭痛學會的會員通知事項,及頭痛相關文章。本園地公開,竭誠歡迎所有頭痛相關醫學著述、病例討論、文獻推介、研討會講座等投稿,稿酬從優。敬請不吝指教,感謝您的支持!

聯絡人:秘書 陳雅惠 會址:台北市士林區文昌路95號B4神經科辦公室 TEL:02-28332211轉2598 FAX:02-28344906

E-MAIL:taiwan.head@msa.hinet.net <http://www.taiwanheadache.com.tw/>