J Oral Pathol Med (2005) 34: 621–5
© Blackwell Munksgaard 2005 · All rights reserved
www.blackwellmunksgaard.com/jopm

CASE REPORT

Non-syndromic multiple supernumerary teeth transmitted as an autosomal dominant trait

P. Batra[1], R. Duggal[1], H. Parkash[2]

[1]*Division of Orthodontics, Department of Dental Surgery, All India Institute of Medical Sciences;* [2]*Department of Dental Surgery, All India Institute of Medical Sciences, New Delhi, India*

Supernumerary teeth are common in the general population and occur more frequently in-patients with family history of such teeth. Multiple supernumerary teeth are associated with cleidocranial dyplasia and Gardner syndrome. However it is rare to find multiple supernumeraries in individuals with no other associated disease or syndrome. We describe the occurrence of multiple supernumerary teeth in a family occurring as a non-syndromal trait. The autosomal dominant transmission of non-syndromal multiple supernumerary teeth is new.
J Oral Pathol Med (2005) **34**: 621–5

Keywords: autosomal dominant transmission; cleidocranial dyplasia; Gardner syndrome; supernumary teeth

Introduction

Supernumerary teeth mean teeth more than the normal number of the full complement of teeth in the primary or permanent dentition (1). They are twice more common in the permanent than in the primary dentition (2). Supernumerary teeth may occur singly, multiply, unilaterally or bilaterally in the maxilla, mandible or both. Their shape and size may resemble the group of teeth at the site where they are found in the jaws or there may be little or no resemblance at all (3).

The prevalence of supernumerary teeth on a population basis ranges from 0.1 to 3.6% (4, 5). The anterior maxillary region appears to be the site of predilection and there is a 10-fold frequency of occurrence in the maxilla (6). The second most frequent site is the distal of the maxillary third molar (6). They occur more frequently in males with the male/female ratio of 2:1 (7).

Supernumerary teeth may erupt normally, stay impacted, appear inverted or assume an ectopic position or an abnormal path of occurrence. Their development might precipitate a variety of complications such as crowding, delayed eruption, diastema development, cystic lesions and resorption of adjacent teeth. Therefore early diagnosis, proper evaluation and appropriate treatment are essential (7).

Multiple supernumerary teeth are associated with cleidocranial dysplasia, and Gardner syndrome. However it is rare to find multiple supernumeraries in individuals with no other associated disease or syndrome (5). A high occurrence rate of 21.2% has been reported in Gardner syndrome, whereas the percentage occurrence in cleidocranial dysplasia is 22% in the maxillary incisor region and 5% in the molar region (8–10).

The cases of non-syndromal multiple supernumerary teeth (11–26) (more than five) where the exact number were stated are listed in Table 1. We describe the occurrence of multiple supernumerary teeth in a family occurring as a non-syndromal trait.

Case report

A 17-year-old girl came to the orthodontic clinic with a chief complaint of forwardly placed front teeth. Intra oral examination revealed retained deciduous teeth namely upper canines, lower left canines and first and second molars as well as lower left canine and first deciduous molar. There was no relevant medical history and the patient was otherwise healthy. Routine radiographic investigations were carried out to see the status of the permanent teeth as well as to evaluate the case orthodontically. The intraoral and extraoral photographs were taken along with the study models (Figs 1 and 2).

The panoramic radiograph however revealed the presence of a number of supernumerary teeth along with the impacted permanent succedaneous teeth (Fig. 3). In the maxilla there were three supernumeraries (one mesiodens and two premolars one on the left and one on the right side). The permanent canines on both the sides were impacted. In the mandible eight supernumeraries were present (two in the incisor region and three supernumerary premolars on both the right and the left sides. Besides the lower canines

Correspondence: Dr Ritu Duggal, Associate Professor, Department of Dental Surgery, All India Institute of Medical Sciences, New Delhi, India. Tel.: 011 659 3231.
Accepted for publication June 22, 2004

Table 1 List of the cases of non-syndromal multiple supernumerary teeth (more than five) reported in literature

No.	*Author/year*	*Age/sex*	*Number of patients*	*Location*
1	Smith 1969 (27)	21/M	19	Not specified
2	Foley and Del Rio 1970 (28)	22/M	14	Not specified
3	Nelson 1971 (11)	22/M	7	Not specified
4	Shapira 1973 (12)	29/M	6	Lower premolars
5	Barnett 1974 (13)	12/M	6	Two upper and lower molars, two lower premolars
6	Finkel et al. 1974 (14)	24/M	9	Two upper molars, one lower molar, five lower premolars and one upper premolar
7	Stevenson and McKechnie 1975 (15)	10/M	11	Five upper anteriors, five lower premolars, one upper premolar
8	Fitzgerald 1978 (16)	20/M	16	Not specified
9	Shusterman et al. 1978 (17)	11/M	8	Not specified
10	Shusterman et al. 1978 (17)	7/F	6	Two upper premolars, two upper and lower anteriors
11	Mercuri and O'Neil 1980 (18)	17/F	27	Not specified
12	Mercuri and O'Neil 1980 (18)	15/F	17	Not specified
13	Rechart 1982 (19)	18/M	7	Four upper premolars, three lower premolars
14	Leslie 1984 (20)	25/M	6	One upper molar, two lower molars, one upper premolar, two lower premolars
15	Acton 1987 (2)	24/M	7	Four upper molars, two lower molars, one lower premolar
16	Yusof and Awang 1990 (22)	24/M	16	Two upper molars, three lower molars, eight lower premolars, three upper premolars
17	Yusof and Awang 1990 (22)	23/F	10	Two upper molars, one lower molar, six lower premolars, one upper premolar
18	Yucel 1992 (23)	22/M	6	Five lower premolars, one upper premolar
19	Hopcraft 1998 (24)	18/M	10	Three maxillary premolars, five mandibular premolars, two upper molars
20	Desai and Shah 1998 (3)	25/M	7	Three upper molars, two upper premolars, two lower premolars
21	Desai and Shah 1998 (3)	36/M	16	Two lower molar, five lower premolars, five upper molars, one upper premolar, three upper anterior
22	Sg'ang'a et al. 2002 (25)	14/M	8	Four upper and four lower premolars
23	Sg'ang'a et al. 2002 (25)	13/F	7	Two upper molars and five lower premolars
24	Umweni and Osunbor 2002 (26)	10 males; three females		Described 13 cases in Nigeria

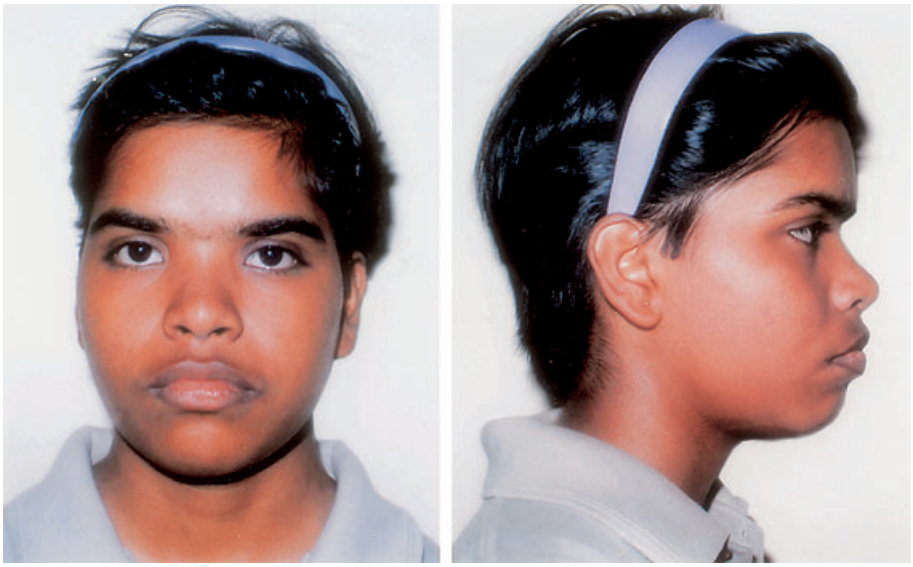

Figure 1 Extraoral photographs of the patient.

and first premolars were impacted on both the sides as well as the left lower second premolar. In view of the multiple supernumeraries the patient was investigated for any feature suggestive of cleidocranial dysplasia and Gardner syndrome. As the patients were normal in her facial appearances, showed no signs of mental retardation and did not exhibit any physical or skeletal abnormality, most of the syndromic conditions

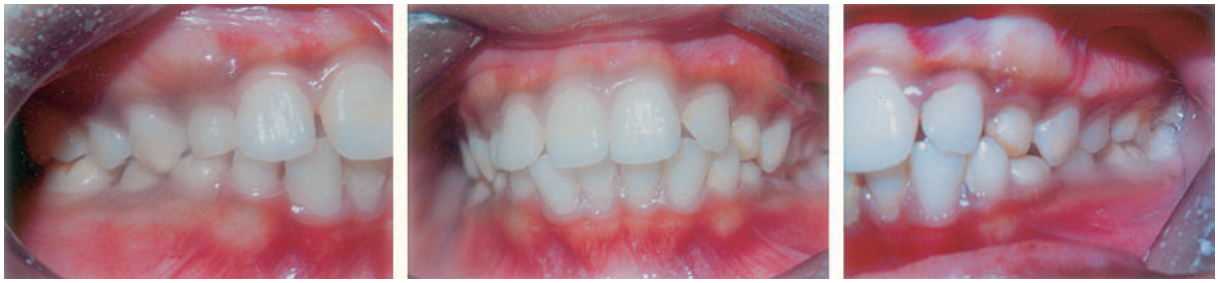

Figure 2 Intraoral photographs of the patient.

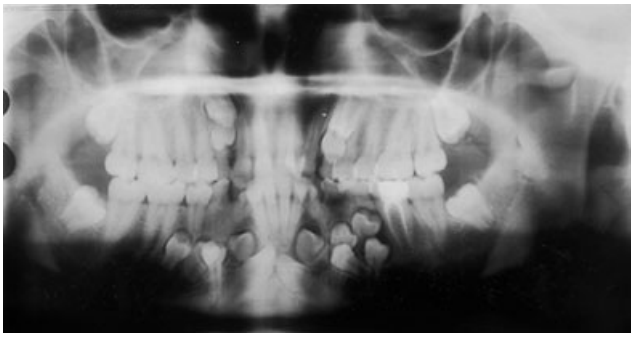

Figure 3 OPG of the patient.

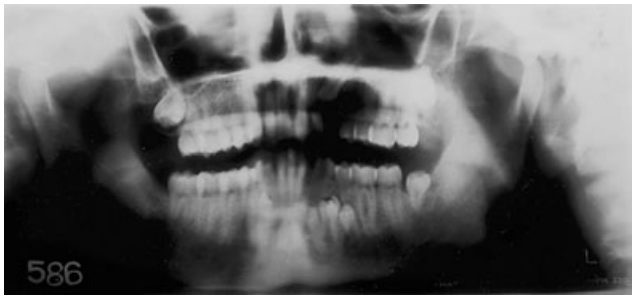

Figure 4 OPG of the patient's brother.

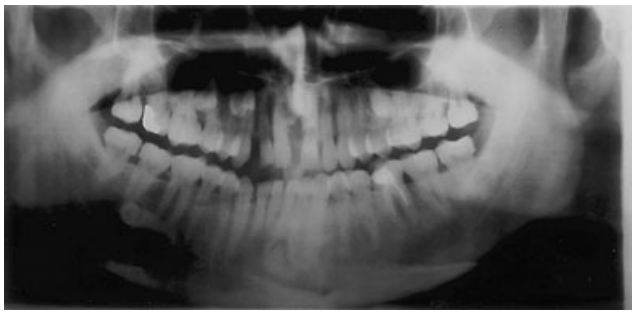

Figure 5 OPG of the patient's father.

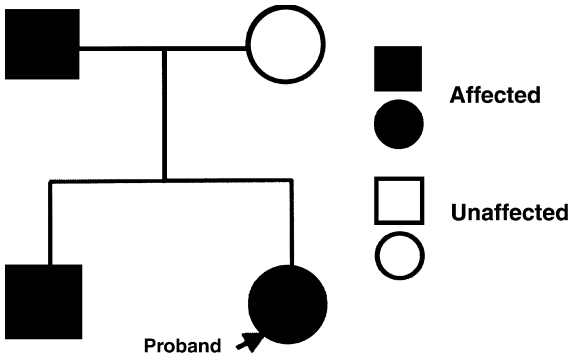

Figure 6 Pedigree showing the autosomal dominant transmission of the multiple supernumerary teeth in the proband and her brother.

were over ruled. This resulted in the dental formula of 43 teeth.

The patient's parents and brother were advised panoramic radiographs to rule out for any supernumeraries. The family of the patient was reluctant to visit our orthodontic clinic. However they got the OPG's (Orthopantomograms) performed from a radiographic center and sent them when the patient reported for their next appointment. The OPG of the patient's elder brother (20 years old) revealed presence of an impacted left upper canine (Fig. 4). Besides supernumerary impacted teeth were present in the eight in number (four incisors and four premolars). The panoramic view of the father revealed the presence of four supernumerary premolars one in the mandible and three in the maxilla (Fig. 5). However no supernumerary tooth was present in the mother's dentition. The pedigree is shown in Fig. 6 and the distribution of supernumerary teeth is given in Table 2.

Discussion

Several theories have been suggested for the occurrence of supernumerary teeth, from the phenomenon of atavism (27) to abnormal activity of the dental lamina, which results in the formation of extra tooth buds by additional formation or splitting of the existing ones (28). Authors have suggested the possibility of a supernumerary tooth arising from clumps of remaining epithelium after the breaking up of the tooth band and becoming activated to tooth formation (dichotomy theory) (29). The association of heredity has also been suggested and most cases are determined by multifactorial inheritance (30).

Table 2 Distribution of supernumeraries in the proband, brother and father

	Molar region (right side)	*Canine-premolar region (right side)*	*Incisor region (right)*	*Incisor region (left)*	*Canine-premolar region (left side)*	*Molar region (left side)*
Proband (total = 11)						
Maxilla		1	1 mesiodens		1	
Mandible		3	1	1	3	
Brother (total = 8)						
Maxilla						
Mandible		2	2	2	2	
Father (total = 4)						
Maxilla		2			1	
Mandible	1					

The cases of non-syndromal multiple supernumerary teeth are more likely to occur in-patients, whose relatives also possessed supernumeraries, although inheritance does not follow a simple Mendelian pattern (7). However the occurrence of multiple supernumeraries in this family suggests that there was an autosomal dominant pattern of inheritance. The case reported above are also unusual in the sense that transmission of multiple supernumerary teeth as an autosomal dominant trait has never been reported before and the peculiar distribution of most of the supernumeraries in the premolar region. In view of the possible sequel associated with these teeth most authors favor surgical removal, although this may cause damage to the adjacent structures, particularly in the mandibular premolar region (31–33).

Gardner syndrome is characterized by the occurrence of multiple impacted supernumerary teeth, which may lead to its early diagnosis (34). This is important as the syndrome consists of multiple polyposis of the large intestine, osteomata of the long bones, skulls and the jaws, sebaceous cysts of the skin and occasional desmoid tumors. It is the intestinal polyps, which are cause for concern, as they might appear later than other symptoms and malignant changes in them are common (35).

Multiple supernumeraries are also found in cleidocranial dysplasia (36). However the other signs of the condition are more likely to be noticed early and are less sinister in their implications than those in Gardner syndrome. These include abnormalities of the skull, jaws and shoulder girdle as well as defects of the vertebral column, pelvis and digits.

We emphasize that the complete medical history is critical when we come across a patient with multiple supernumeraries. One has to rule out all those medical syndromes associated with them before labeling it as a case of non-syndromic multiple supernumerary teeth.

References

1. Gorlin RJ, Goldman HM. *Thoma's oral pathology*. 6th edn. St Louis: C V Mosby Company, 1970.
2. Clayton JM. Congenital dental anomalies occurring in 3557 children. *ASDC J Dent Child* 1956; **23**: 206–8.
3. Desai RS, Shah NP. Multiple supernumerary teeth in two brothers: a case report. *J Oral Pathol Med* 1998; **27**: 411–3.
4. Schulze C. Incidence of supernumerary teeth. *Dent Abstr* 1961; **6**: 23.
5. Yusof WZ. Non syndrome multiple supernumerary teeth: literature review. *J Can Dent Assoc* 1990; **56**: 147–9.
6. Nazif MN, Ruffalo RC, Zullo T. Impacted supernumerary teeth: a survey of 50 cases. *J Am Dent Assoc* 1983; **106**: 201–4.
7. Brook AH. A unifying aetiological explanation for the tooth number and size. *Arch Oral Biol* 1984; **29**: 373–8.
8. Ida M, Nakamura T, Utsunomiya J. Osteomatous changes and tooth abnormalities found in the jaws of patients with adenoma coli. *Oral Surg* 1981; **52**: 2–11.
9. Shafer WG, Hine MK, Levi BM. *Textbook of oral pathology*. 4th edn. Philadelphia: WB Saunders Co, 1983.
10. Jensen HL, Kreiborg S. Development of dentition in cleidocranial dysplasia. *J Oral Pathol Med* 1990; **19**: 89–93.
11. Nelson JF. Multiple supernumerary premolars and incisors. *Oral Surg* 1971; **31**: 279–80.
12. Shapira Y. Six supernumerary mandibular premolars. *J Dent* 1973; **1**: 263–4.
13. Barnett BS. A case of multiple supernumerary premolars. *Br J Orthod* 1974; **1**: 217–8.
14. Finkel AN, Solondz G, Friedman J. Multiple supernumerary and impacted teeth. *Oral Surg* 1974; **37**: 976–7.
15. Stevenson W, McKechnie AD. Recurring supernumerary teeth. Report of a case. *Oral Surg* 1975; **40**: 76–80.
16. Fitzgerald BE, Zallen RD. Multiple impacted supernumerary teeth. *Oral Surg Oral Med Oral Pathol* 1978; **46**: 733.
17. Shusterman S, Hertzberg J, Hilzenrath S. Multiple supernumerary teeth: report of two cases. *ASDC J Dent Child* 1978; **45**: 469–72.
18. Mercuri LG, O'Neill R. Multiple impacted and supernumerary teeth in sisters. *Oral Surg Oral Med Oral Pathol* 1980; **50**: 293.
19. Reichart PA. Duplication of all permanent premolars. *Oral Surg* 1982; **53**: 437.
20. Leslie JC. Multiple supernumerary teeth. *Oral Surg* 1984; **57**: 463.
21. Acton CH. Multiple supernumerary teeth and possible implications. *Aust Dent J* 1987; **32**: 48–9.
22. Yusof WZ, Awang MN. Multiple impacted supernumerary teeth. *Oral Surg Oral Med Oral Pathol* 1990; **70**: 126.
23. Yucel E. Multiple supplemental premolars. *Oral Surg Oral Med Oral Pathol* 1992; **74**: 384.
24. Hopcraft M. Multiple supernumerary teeth. Case report. *Aust Dent J* 1998; **43**: 17–9.
25. Sg'ang'a PM, Guthua SW, Ng'ang'a RN. Multiple supernumerary teeth in association with malocclusion: a report of two cases. *East Afr Med J* 2002; **79**: 221–3.

26. Umweni AA, Osunbor GE. Non syndrome multiple supernumerary teeth in Nigerians. *Odontostomatol Trop* 2002; **25**: 43–8.
27. Smith JD. Hyperdontia: report of a case. *JADA* 1969; **79**: 1191–2.
28. Foley MF, Del Rio CE. Supernumerary teeth: report of a case. *Oral Surg* 1970; **30**: 60–3.
29. Saarenmaa L. The origin of supernumerary teeth. *Acta Odontol Scand* 1950; **9**: 293–303.
30. Farmer ED, Lawson FE. *Stones oral and dental diseases*. 5th edn. Edinburgh and London: E&S Livingstone Ltd, 1966.
31. King NM, Lee AMP, Wan PKC. Multiple supernumerary premolars: their occurrence in three patients. *Aust Dent J* 1993; **38**: 11–16.
32. Zhu JF, Marcuchamer M, Kink DL, Henry RJ. Supernumerary and congenitally absent teeth: a literature review. *J Clin Pediatr Dent* 1996; **20**: 87–95.
33. Bodin I, Julin P, Thompson M. Hyperdontia: VI. Supernumerary premolars. *Dentomaxillofac Radiol* 1981; **10**: 99–103.
34. McFarland PH, Sheetz WL, Knisley RE. Gardners syndromer: report of two families. *J Oral Surg* 1968; **26**: 632–8.
35. Fader M, Kline SN, Spatz SS, Zurbrow HJ. Gardners syndrome (intestinal polyposis, osteomas, sebaceous cyst) a new dental discovery. *Oral Surg* 1962; **15**: 153–72.
36. Atasu M, Dumlu A, Ozbayrak S. Multiple supernumerary teeth in association with cleidocranial dysplasia. *J Clin Pediatr Dent* 1996; **21**: 85–91.

This document is a scanned copy of a printed document. No warranty is given about the accuracy of the copy. Users should refer to the original published version of the material.